Original article

Type I spinal arteriovenous fistulae exception: A case for reclassification

Une forme exceptionnelle de fistule artério-veineuse rachidienne de type I : un cas justifiant une nouvelle classification

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ABSTRACT

Objective. – Type I spinal arteriovenous lesions represent dorsal dural arteriovenous fistulae with no spinal artery involvement. We report an exception to this and propose dividing Type I lesions into dorsal and ventral categories.

Methods. – A 51-year-old patient presented with a partial Brown-Sequard syndrome. An angiogram revealed a spinal arteriovenous fistula, most prominently being fed by a radicular artery arising from the right vertebral artery with only ventral venous drainage.

Results. – This feeder was selected and embolized with onyx, however residual fistula persisted and the patient subsequently underwent microsurgical disconnection. At six-month follow-up, patient motor deficits have resolved and difficulty with proprioception is improving.

Conclusion. – Type I dural arteriovenous fistulae are associated with dorsal venous drainage. Our case demonstrates an exception to this. Recognizing this exception was crucial, because it allowed for simple microsurgical disconnection. We propose that type I dural arteriovenous fistulae be subdivided into ventral and dorsal on the basis of drainage pattern. This differentiation is critical because lesions with ventral drainage have traditionally been classified as type IV. These lesions have a different treatment method given involvement of spinal arteries.

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RéSUMÉ

Objectif. – Le type I des lésions artério-veineuses rachidiennes désigne les fistules artério-veineuses durales dorsales sans participation de l’artère spinales. Nous rapportons une exception à cette définition et proposons de diviser les lésions de type I en deux sous-catégories : lésions dorsales et lésions ventrales.

Méthodes. – Un homme de 51 ans était pris en charge pour un syndrome de Brown-Séquard partiel. L’angiographie a révélé une fistule artério-veineuse rachidienne principalement alimentée par une artère radiculaire issue de l’artère vertébrale droite, avec un drainage veineux uniquement ventral.

Résultats. – Le vaisseau a été cathétérisé et embolisé à l’onyx. Une fistule résiduelle a cependant persisté justifiant d’une procédure de déconnexion microchirurgicale. À six mois postopératoires, le déficit moteur du patient a régressé et les troubles de la proprioception sont en cours d’amélioration.

Conclusion. – Les fistules artério-veineuses dorsales de type I sont associées à un drainage veineux dorsal. Notre cas illustre une exception à cette définition. Avoir reconnu cette exception est primordial, car cela a permis une simple déconnexion microchirurgicale de cette fistule. Nous proposons que les fistules artério-veineuses dorsales de type I soient subdivisées en fonction de la position ventrale ou dorsale de leur drainage. Cette différenciation est essentielle car les lésions à drainage ventral ont traditionnellement été considérées comme des lésions de type IV qui ont un traitement différent compte tenu de l’implication des artères spinales.

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1. Introduction

In 1914, Dr. Charles Elsberg performed the first successful operation for a spinal arteriovenous malformation (AVM) [1]. This ultimately heralded an age in the 1960s when improved selective spinal angiography increased our understanding of these relatively rare lesions and lead to an initial classification scheme that divided spinal AVMs into three types:

- type I dorsal extramedullary;
- type II intramedullary AVM with multiple feeders;
- type III, extensive juvenile malformation [2–6].

In 1986, Heros et al. added a Type IV AVM, specifically one that was in the ventral spinal cord and had a fistulous connection with the anterior spinal artery [7,8]. In 1992, Anson and Spetzler summarized the classification scheme and proposed treatment paradigms associated with each subtype of AVM [9]. This classification, along with the subsequent modification, has become the most widely accepted in clinical practice [8].

In this report, we describe a patient with a spinal arteriovenous fistula with supply from the ventral radiculomeningeal branch that does not fit into the existing classification scheme. Adequately classifying such a lesion has significant treatment implications because there is no involvement of either the anterior or posterior spinal arteries. The four type classification scheme that was proposed by Anson and Spetzler remains the most popular and widely utilized grading scheme for these lesions and it is utilized to guide treatment. We report on a patient that does not fit into this popular classification scheme. We propose that Type I spinal AVMs be sub divided into dorsal extramedullary lesions and a new category, ventral extramedullary lesions. The latter is distinct from type IV lesions because there is no spinal artery involvement. We feel that it is important to differentiate these lesions because in the proposed type I ventral extramedullary lesion, the fistula point is in the ventral nerve root sleeve and no spinal artery involvement is present. This has significant treatment implications.

1.1. Case report

A 51-year-old Caucasian female with a past medical history significant for hypothyroidism presented to our institution for evaluation of progressive numbness, weakness and difficulty with ambulation. Patient reported the onset of neck pain and left arm pain 2 months earlier. On exam she exhibited a partial Brown-Séquard syndrome. Her exam was significant for 4-/5 left triceps weakness, 3/5 left hand intrinsic muscle weakness and 4+/5 weakness throughout the left lower extremity. The patient’s sensory exam was notable for decreased proprioception on the left side and decreased pain and touch involving the right leg. She displayed a Hoffman’s reflex, as well as, clonus on the left. CT-angio of the neck revealed a vascular lesion at the level of C6 which was ventral to the spinal cord (Fig. 1). MRI demonstrated a 1.1 cm vascular structure, ventral to the spinal cord, compressing the left side of the spinal cord at the level of C6. Associated with this structure were multiple serpiginous tubular structures consistent with engorged veins. Increased T2 signal was present from the cervicomedullary junction down to the mid-thoracic region (Fig. 2).

An angiogram revealed a spinal arteriovenous fistula, most prominently being fed a radicular artery arising from the right vertebral artery (Fig. 3). Visualization of the adjacent levels confirmed contribution from the level above the lesion to the anterior spinal artery (ASA). Angiography also revealed the pathology not to involve the ASA. Superselective angiography was performed. This feeder was embolized using Onyx. After embolization, a feeder from the thyrocervical trunk was revealed. This feeder was selected and embolized with onyx. Injection of the right subclavian artery revealed persistent, but much less robust, filling of the spinal AVM (Fig. 4). Injection of the thyrocervical and costocervical trunks suggested arterial contribution from both these arteries. No arterial feeder was able to be isolated at this point. The patient returned to the ICU. Following the intervention, the patient’s neurological exam remained stable.

A CT-angio of the neck revealed persistent filling of the previously identified ventral venous varix arising at the C6 vertebral level, although the varix was much diminished in appearance (Fig. 5).

Patient was then taken to the operating room. Neuromonitoring was used. She was placed in a Mayfield headrest then turned prone. Under fluoroscopic guidance the C6-7 vertebral level was easily identified using the previously placed Onyx as a landmark (Fig. 6). The site was confirmed with traditional methods. C5-C7 laminectomies, a partial right C5-6 facetectomy and a durotomy were performed to expose the right-sided C7 nerve root. On the

![Fig. 1. Sagittal and coronal CT-angio reconstructions demonstrate serpiginous vessels and venous varix ventral to the spinal cord.](image-url)
ventral nerve root an arterialized vein was encountered. We also encountered an arterialized vein ventral to the spinal cord. Arterial flow was confirmed with Doppler ultrasound. A temporary clip was placed on the lesion near the nerve root. Arterial flow, confirmed by Doppler ultrasound, was obliterated distal to the clip and in the arterialized vein which was ventral to the spinal cord. The temporary clip was left in place for 20 minutes without observed changes in neuromonitoring. A permanent clip was used to occlude the fistula. The wound was closed in a standard fashion. Postoperatively the patient's neurologic exam was stable compared to preop. CT-angiography performed on postoperative day 1 did not reveal any filling of the AVM (Fig. 7). On postoperative day 2 she was discharged to a rehabilitation center.

At her six-month outpatient follow-up visit, patient had regained full strength of her lower extremities and had improving proprioception.

2. Discussion

There is no doubt that spinal AVMs are rare lesions, however with increasing experience with these lesions, subtleties can now be appreciated that have significant treatment implications. In this report we demonstrate a spinal AVM that did not fit into the traditional classification scheme. Based on the ventral drainage and absence of an intramedullary nidus, our reported AVM was initially considered to be a type IV AVM. It does not, however, fit...
into this category because there is no contribution from the intrinsic blood supply of the spinal cord, specifically the anterior spinal artery. It did not fit into the type I AVM because it was not dorsal extramedullary (Fig. 8), however its treatment most closely resembled this AVM because the fistula point was at the dural root sleeve. The venous drainage of the reported lesion was exclusively ventral perimedullary. We therefore propose subdividing type I AVM into dorsal extramedullary and a new category ventral extramedullary (Fig. 9). In order for a lesion to be classified as Type I extramedullary, spinal angiography must confirm absence of anterior spinal artery involvement. Given that the fistula point in these lesions is in the dural root sleeve and supplied by a radiculomeningeal artery, a dorsal approach with far lateral extension may be sufficient to disconnect the fistula.

Type I AVMs have most commonly been referred to as dural arteriovenous fistulas. Prior to the introduction of our proposed new subclassification of this group, they were also known as dorsal AVMs [5,9]. They are primarily found in the thoracic spinal cord and conus medullaris. Their hallmark is that they consist of either a single or multiple arterial feeders that enter the dura dorsolaterally at the dural root sleeve. The venous outflow of the fistula drains intradurally into an arterialized vein which runs along the dorsal surface of the spinal cord [9]. These lesions cause symptoms by progressive venous hypertension with chronic intramedullary ischemic changes. These lesions most commonly affect males between the fifth and eighth decades causing a radiculomyelopathy with progressive neurological deterioration [10]. Good results are obtained with obliteration of the fistula [11–15].

As endovascular treatment options have increased, there are some centers that now advocate it as first line treatment for type 1 lesions [16]. There is uniform agreement that in order for this strategy to work, the distal feeding vessel and the proximal portion of the efferent intradural arterialized veins need to be occluded [16–19]. Unfortunately, follow-up results show high recanalization rates and complete obliteration of the fistula being less than 25% [16,17,19]. Microsurgical treatment to disconnect the fistula thus remains the treatment of choice for these lesions.

Type IV spinal AVMs were first described by Djindjian et al. in 1977 and subsequently added by Heros et al. to the classification scheme in 1986 [7,20]. These lesions are most often anterior or anteriorlateral to the spinal cord and are extramedullary. They are usually supplied by the anterior spinal artery, however there can be posterior locations supplied by the posterior spinal arteries [9]. These lesions most often occur at the conus medullaris. They have further been subdivided into three subtypes with treatment implications [9,18]. What is consistent among all these subtypes, however is involvement of an intrinsic spinal cord artery, usually the anterior spinal artery.

In conclusion, based on the unique anatomical findings of our reported lesion, we propose subdividing type 1 AVMs into dorsal extramedullary and a new category, ventral extramedullary. These lesions are supplied by radiculomeningeal arteries with fistulous connection with intradural arterialized venous drainage.
without any involvement of the ASA. The type I AVM ventral extramedullary subtype can be confused with Type IV, however the distinguishing feature is that the former has no intrinsic spinal cord artery involvement. Since the fistula point is in the dural root sleeve, a dorsal approach with a far lateral extension may be sufficient to disconnect the fistula in the type I ventral extramedullary lesion.

Disclosure of interest

The authors have not supplied their declaration of conflict of interest.

References


