Extradural arteriovenous fistulas involving the vertebral artery in neurofibromatosis Type 1

Case report

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✓ Spinal extradural arteriovenous fistulas (AVFs) are rare lesions that may be associated with neurofibromatosis Type 1 (NF1). In these patients, the shunt typically occurs between the V2 segment of the vertebral artery and the epidural venous plexus. Previously, reported cases have been treated either by using endovascular embolization or, sporadically, by open surgery. In surgical reports, proximal deafferentation or manipulation of the venous portion of the shunt—including suture, resection, or open embolization of the epidural ectasia—was attempted with variable results. The authors report on a case of a young patient with NF1 who underwent emergency surgical disconnection of a cervical extradural AVF after previously unsuccessful endovascular and surgical therapy. The lesion drained into a giant intrathecal varix, causing severe myelopathy. After surgery, the patient recovered almost completely. This experience clarified the surgical anatomy of these malformations and showed that, when surgery is necessary, the optimal treatment providing complete and permanent cure of this condition is direct closure of the epidural shunt pedicle. (DOI: 10.3171/SPI/2008/8/2/181)

KEY WORDS • extradural arteriovenous fistula • neurofibromatosis • vertebral artery

Extradural AVFs involving the V2 segment of the VA are rare lesions, reportedly associated with NF1. These malformations may drain within the prevertebral venous system or intrathecally through the epidural venous plexus. Endovascular embolization is the treatment of choice for these lesions.5,6,14 In some cases endovascular obliteration of the shunt can be difficult to perform,3 and surgical treatment has been deemed hazardous or even impossible.1,3,13,15,16 The few reported cases involved, with variable results, direct manipulation of the venous compartment of the shunt through suture,7,13 or open embolization3 of the epidural ectasia. We report the case of a young patient with NF1 who underwent surgical obliteration of a cervical extradural AVF that was draining into a giant intrathecal varix, using direct clip placement for the epidural shunt. The aim of this report is to illustrate the anatomical and surgical findings in a paradigmatic case.

Case Report

This 26-year-old man with NF1 sustained a mild whiplash injury. A few months later he developed sensory disturbances and progressive weakness of his 4 limbs. A cervical MR image disclosed a vascular lesion compressing the spinal cord at C2–3 (Fig. 1). Use of DSA revealed an arteriovenous shunt between multiple arteries of the neck and the intrathecal venous system. A hypertrophic, tortuous left VA was the major feeder of the malformation (Fig. 2). Elsewhere the patient had undergone unsuccessful endovascular balloon occlusion of the shunt pedicle. Each attempt—performed with clinical monitoring—was stopped because of immediate worsening of the tetraparesis. Surgery was also attempted without success. Eventually, coil
embolization of feeding arteries arising from the external carotid artery and the deep cervical artery was performed. One month later the patient was admitted to our institute with complete tetraparesis, dysphagia, and respiratory failure. Repeated DSA showed further recruitment of arterial feeders and enlargement of the intrathecal drainage. Computed tomographic angiography showed the broad and apparently single shunt pedicle within the C2–3 neuroforamen (Fig. 3). Emergency surgery was performed.

With the patient supine, arterial deafferentation was performed by proximal ligation of the first (V1) segment of the VA, the thyrocervical trunk, and the external carotid artery. Intraoperative angiography confirmed flow reduction through the malformation, although retrograde filling by the distal VA was still evident. A posterior approach was then performed. On opening the dura, the spinal cord was noted to be severely compressed by a bulging dural wall (Fig. 4) that was temporarily shrunk using long aneurysm clips. The VA was trapped by clipping the V4 segment. Attention was then directed to the left C2–3 foramen, which was eroded by a pinkish mass (the shunt pedicle), resembling a swollen C-3 nerve root. Due to its size, the pedicle could not be dissected circumferentially. Any attempt to blindly clip the shunt pedicle close to the dural sac resulted in profuse bleeding.

Intraoperative angiography showed that the left VA, despite being trapped, was filled by the contralateral VA through cross-anastomoses (Fig. 5). A different strategy was then adopted: the transverse foramen of C-2 was exposed and drilled to dissect the VA free. The artery was then followed proximally, encountering on its medial side a wide-necked anastomosis with the arterialized epidural plexus. The anastomosis was closed using an aneurysm clip, and DSA showed complete disappearance of the shunt (Fig. 6). An MR image obtained 1 month after surgery showed that the spinal cord had regained a normal shape (Fig. 7). One year after surgery, computed tomography angiography confirmed obliteration of the fistula. The patient’s neurological examination was normal, except for minimal weakness of the left hand.

Discussion

Extradural AVFs associated with NF1 have been documented in the literature.1–4,6,8–10,12–16 Our experience, including surgical inspection of the lesion, corroborated some conclusions drawn from the study of Hauck and Nauta: 1) the fistula is located extradurally and, therefore, is only apparently similar to the more common dural fistulas of the spine; 2) an unpredictable number of arteries converge at the VA, which remains the only direct feeder of the shunt; and 3) the fistula consists of a single-hole, high-flow anas-

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**Fig. 1.** Sagittal T1-weighted MR image showing a large lesion effacing the spinal canal at C2–3.

**Fig. 2.** Images obtained using DSA. Injection of the left VA (A) shows a fistulous connection between the artery and enlarged intrathecal veins. A large venous ectasia is evident. Injection of the external carotid artery (B) and thyrocervical trunk (C) also demonstrates the shunt.
tomosis between the VA and the epidural venous plexus (Fig. 8). These observations may explain why the ideal route for endovascular occlusion of the shunt is usually the VA itself. This arterial approach may be infeasible in the presence of some factors, such as vessel tortuosity, previous occlusion of the VA, or failure to tolerate sacrifice of the VA if selective closure of the shunt pedicle cannot be performed. In our case, balloon obliteration of the shunt was attempted but resulted in neurological worsening. The mechanism underlying this event is unclear, but inadvertent occlusion of radiculomedullary arteries or poor compliance with the flow redistribution following abrupt closure of the shunt may be two possible explanations.

The decision to treat this lesion surgically was associated with some concerns. The optimal strategy is not defined by the available literature. Clipping, resection, or direct embolization of the venous ectasia have been attempted as the
sole treatment method, with variable results. In general, the effectiveness and durability of these approaches are unclear because they leave the core of the fistula intact.

Direct obliteration of the vertebral–epidural anastomosis should be the therapy of choice and, to our knowledge, has not been illustrated previously.

From a technical viewpoint, this case added operative insights to the principles previously summarized. The profuse bleeding encountered while trying to approach the venous pedicle suggests that even an extensive arterial deafferentation does not allow safe manipulation of the malformation. The intraoperative angiograms obtained in our case showed that anastomoses that are not apparent initially may manifest after closure of major feeding arteries. Moreover, the arterialized wall of the epidural venous plexus is thicker than usual but still fragile; therefore, dissection of the wall close to the dural sac should be kept to a minimum. Our experience suggests that unroofing the transverse foramen immediately adjacent to the fistula, following the VA, and clipping the abnormal anastomosis on the medial side of the artery is the safest and most logical strategy. This approach keeps the surgeon away from the weakest segment of the fistula and, at the same time, provides a true proximal control of the blood flow. Retrospectively, we are prone to believe that VA sacrifice, obtained in our case as in most other cases successfully treated using endovascular embolization, might be avoided. Laminectomy or laminoplasty do not seem essential, except for emergent decompression of the spinal cord or to verify disconnection of the shunt by direct inspection or puncture of the venous ectasia; intraoperative angiography might serve the latter purpose adequately.

**Conclusions**

Intrathecal extradural AVFs associated with NF1 appear to share typical features. Whenever endovascular occlusion of the shunt cannot be performed, surgical disconnection can be performed with an acceptable margin of safety. The case reported here provides some insights into the surgical anatomy of the vertebral–epidural shunt that could enhance the surgeon’s confidence when approaching these difficult lesions.

**References**

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