

# Endovascular embolisation of a cervical spinal AVF in a patient with NF1

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## Abstract

Vascular abnormalities associated with neurofibromatosis type 1 (NF1) are well described. Spinal arterio-venous fistula (AVF) is a rare finding in NF1 and may present with neurological symptoms that require treatment. Management of spinal AVFs can be endovascular or surgical. We present a patient with known NF1 and neurological symptoms due to a spinal AVF demonstrated by imaging, who required a combination of endovascular and surgical management.

## History

A 38-year-old woman with known NF1 presented with right-sided 12th cranial nerve palsy. On examination, she also had a continuous bruit over the right carotid space.

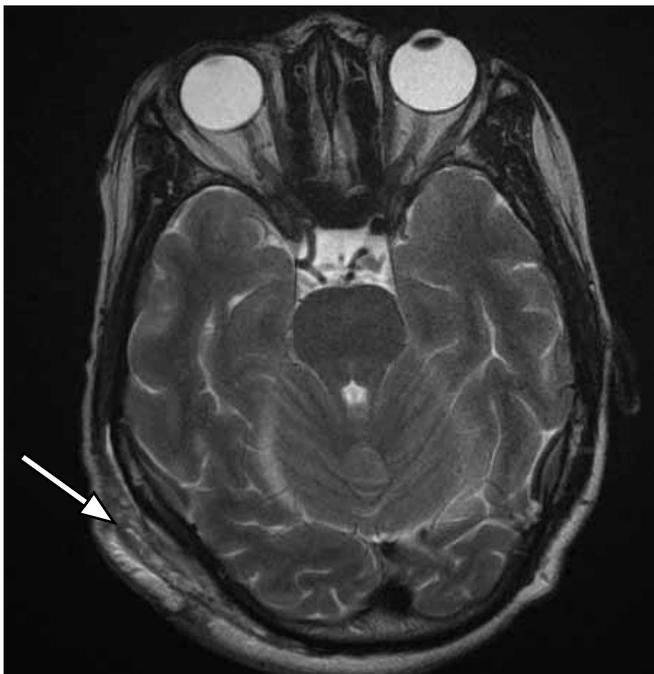


Fig. 1. Axial T2-weighted image of the brain demonstrating the right occipital plexiform neurofibroma (white arrow).

## Imaging findings

Magnetic resonance imaging (MRI) demonstrated a right-sided occipital plexiform neurofibroma in keeping with NF1 (Fig. 1). MRI of the cervical spine demonstrated a large signal void in keeping with an extradural spinal AVF. There was a significant intra-spinal component causing a compressive myelopathy of the cervical spine (Fig. 2), including the medulla at the level of the right 12th cranial nerve, accounting for the clinical presentation. Magnetic resonance angiography (MRA) demonstrated the large right-sided cervicle spinal AVF fed by the right vertebral artery and draining into the epidural veins and the right jugular veins (Fig. 3). Digital subtraction angiography (DSA) confirmed the cervical spinal AVF, demonstrating the shunting through an intricate web of communications between the right vertebral artery and drainage into the right epidural and right internal and external jugular veins (Figs 4a and 4b).

## Treatment

Treatment was initially via endovascular means. Owing to the complex anatomy of the spinal AVF, the fistula orifice was not identified.

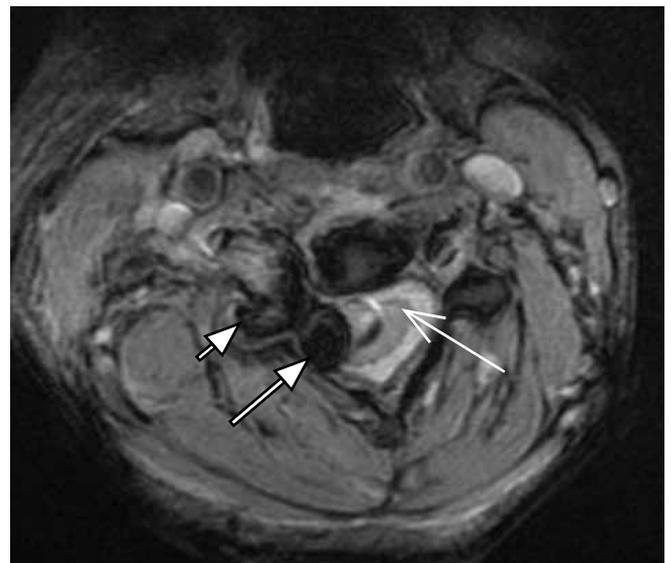


Fig. 2. Axial T2-weighted gradient echo image of the neck demonstrating the large serpiginous signal void (arrow head) of the AVF with an intra-thecal component (filled arrow) causing mass effect on the cervical spinal cord (open arrow).



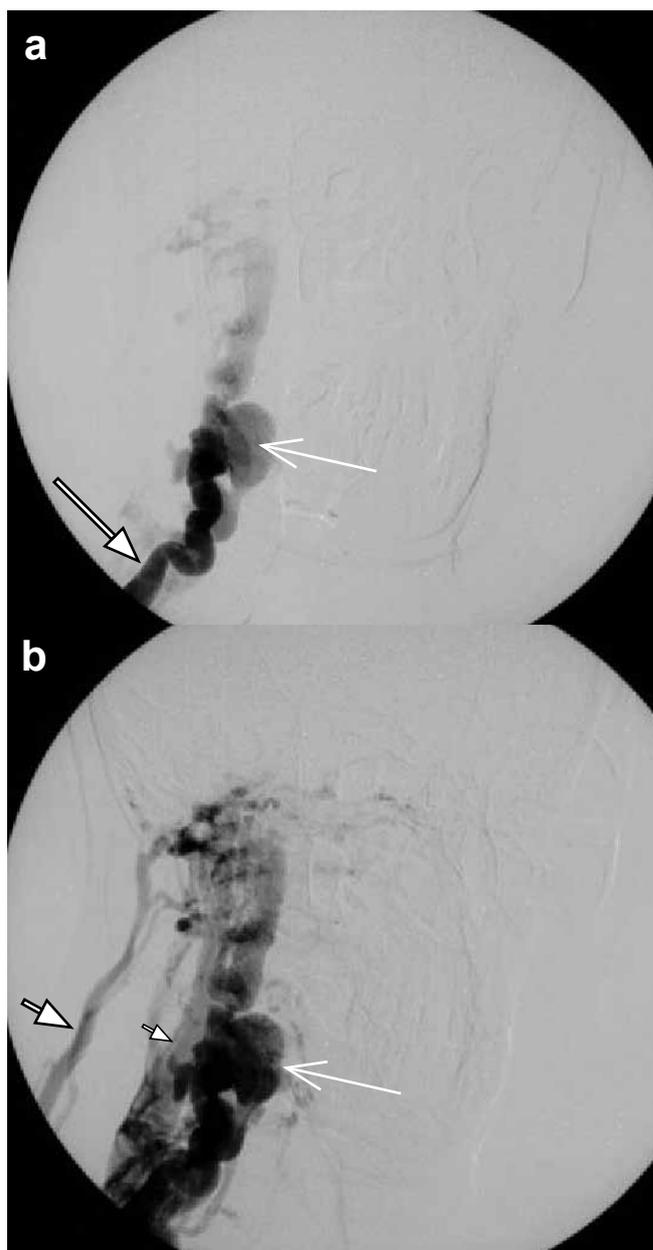
*Fig. 3. MRA demonstrating the cervical spinal AVF fed by the right vertebral artery (arrow head). Note the engorged epidural vein (open arrow) and the right internal jugular vein (filled arrow).*

Therefore, 2 IMWCE 35-6-6 coils were inserted into the AVF from the ipsilateral vertebral artery. As this was unsuccessful in treating the fistula, an Amplatzer vascular plug was placed into the proximal right vertebral artery. However, selective micro-catheterisation of the left vertebral artery demonstrated persistence of the spinal AVF that was now fed by the contralateral vertebral artery (Fig. 5). Selective micro-catheterisation of the left vertebral artery to occlude the distal aspect of the right vertebral artery was attempted but not successful. Therefore, surgical clipping of the distal right vertebral artery at the level of the foramen magnum was performed.

## Discussion

NF1 is one of the most common autosomal-dominant inherited genetic disorders, with an incidence of approximately 1:3 500.<sup>1</sup> Patients with NF1 may present with a spectrum of vascular lesions affecting the central nervous system, including occlusions, hypoplasia of intracranial arteries, 'moyamoya' vessels and aneurysms.<sup>1</sup> Cutaneous neurofibromata, as were present in our patient, are pathognomonic for NF1 and can also be associated with an abnormal vascular supply that may cause increased bleeding during surgical resection.<sup>1</sup>

Spontaneous spinal AVFs associated with NF1 are an uncommon association.<sup>1,2</sup> The combination of skeletal abnormalities of the spine with abnormal vessel fragility in NF1 may predispose patients to the development of spontaneous high-flow spinal AVFs.<sup>1</sup> Ninety-seven per cent of spontaneous spinal AVFs associated with NF1 occur in the cervical spine and arise from the vertebral artery.<sup>1</sup> These fistulae



*Figs 4a and 4b. DSA of the right vertebral artery, confirming the cervical spinal AVF (open arrow), demonstrating the intricate web of communications between the dysplastic right vertebral artery (filled arrow) and the right internal (smaller arrow head) and external jugular (larger arrowhead) veins.*

usually drain into the vertebral venous plexus, or intrathecally via the epidural venous plexus.<sup>2</sup> The clinical presentations of spinal AVFs include radiculomyelopathy (78%), bruit (50%), tinnitus (10%), cranial neuropathy (3%) or a pulsatile neck mass (3%).<sup>1</sup> A cranial neuropathy and bruit were described in our patient. A direct connection between an extradural spinal/radiculomedullary artery and adjacent vein lead to the development of a high-flow fistula with engorgement of the epidural venous system and potential compression of the spinal cord, resulting in progressive myelopathy (Fig. 2).<sup>3</sup> The high venous pressure in the epidural venous system can also lead to intradural venous hypertension by increasing the overall resistance to outflow. The large amount of shunting of arterial blood into the venous system can also steal blood from the spinal cord, leading to a myelopathy.<sup>3</sup>



*Fig. 5. DSA post embolisation with the catheter tip in left (contra-lateral) vertebral artery (open arrow) demonstrates persistence of the spinal AVF, which is fed by the contra-lateral vertebral artery. Note the Amplatzer vascular plug in the proximal right vertebral artery (filled arrows) and the 2 IMWCE 35-6-6 coils (arrowhead) within the spinal AVF.*

Confusion of spinal AVFs with spinal neurofibromata may occur on MRI because of the vascularity of some neurofibromas, as well as the similar appearances of neurofibromata and distended vessels as dumbbell-shaped masses associated with vertebral scalloping. However, a distinct flow-related signal void usually indicates a spinal AVF.<sup>1</sup>

The classification of complex vascular lesions associated with NF1 is controversial. Some authors consider the lesions as spinal arterio-venous malformations (AVMs), while other authors describe them as spinal AVFs.<sup>1</sup> On evaluation of our patient on MRI, MRA and DSA,

the characteristic single vertebral artery that feeds this spinal AV lesion would cause the lesion to be classified as a spinal AVE.<sup>1,3</sup> However, a confounding factor in this classification is the number of engorged and tortuous collaterals and draining vessels that can simulate the multiple feeders of an AVM. Super selective angiography is a valuable investigation in this differentiation.<sup>1</sup>

The goal in managing spinal AVFs in NF1 is complete occlusion of the fistula, i.e. occlusion of the vertebral-epidural anastomosis.<sup>1,2</sup> Extradural spinal AVFs rarely require open surgery as they can be usually be treated effectively by endovascular procedures, which should therefore be first-line management.<sup>1-3</sup>

Endovascular occlusion has a lower risk of bleeding compared with open resection.<sup>1</sup> The endovascular approach involves occlusion of the vertebral artery first distal to the fistula via the ipsilateral vertebral artery, then fistula occlusion and proximal embolisation.<sup>1</sup> Owing to the complex anatomy as described in our patient, the fistula orifice was not identified and the ipsilateral vertebral artery distal to the fistula could not be occluded. Proximal occlusion of the right vertebral artery using the Amplatzer device was successful. Use of a detachable balloon would have been better suited in this case, owing to the tortuosity of the dysplastic right vertebral artery allowing better passage of the delivery system; however, detachable balloons were not available at our institution at the time of this procedure. Using a contra-lateral approach to occlude the fistula distally may be difficult and risky, and in our patient was not successful. A further treatment option is the use of a covered stent to occlude the fistula orifice, while preserving the vessel.<sup>1</sup> The fragility or tortuosity of the vessels in NF1 can further complicate endovascular procedures, and delayed recanalisation may be a problem.<sup>1,2</sup> For large fistulae with multiple collateral feeders, as described in our patient, embolisation alone may be difficult or unsuccessful. Surgery is generally reserved for incomplete endovascular cure and to relieve compression of the dura and spinal cord.<sup>2,3</sup>

## Conclusion

This case highlights the importance of MRA as a sensitive and non-invasive modality in the imaging of NF1 and the diagnosis of cervical spinal AVFs. Spontaneous spinal AVFs from the vertebral artery may simulate a cervical spinal AVM owing to multiple collateral arterial feeders arising from the vertebral artery in NF1. Endovascular techniques is the first-line management for cervical spinal AVFs, but may need to be performed in combination with surgery for large fistulae with multiple collateral feeders, as described in this patient.

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