Endovascular management of spontaneous vertebrovertebral arteriovenous fistula associated with neurofibromatosis 1

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Abstract
Extra cranial vertebrovertebral arteriovenous fistulas (VV AVF) are commonly associated with trauma. Their presentation may vary from palpable thrill and myelopathy or myeloradiculopathy. Sudden onset paraparesis is rare.

Key words: Embolization; neurofibromatosis type 1; NF 1; vertebrovertebral arteriovenous fistula

Introduction
Extracranial vertebrovertebral arteriovenous fistula (VV AVF) is a rare condition. It is commonly caused by trauma, but can be congenital, spontaneous, iatrogenic, idiopathic, or related to dysplastic syndromes. We describe a case of spontaneous VV AVF associated with neurofibromatosis Type 1. The patient presented to us with sudden-onset right arm weakness. He was successfully treated by endovascular embolization.

Case Report

History
A 42-year-old man with neurofibromatosis (NF) Type 1 presented with neck pain, weakness of both the arms, and humming sound on the right side of the neck for past 3 months. The weakness was more on the right arm.

He was admitted for further evaluation and management. His right upper limb power suddenly deteriorated during the hospital stay.

Clinical examination
General examination of the patient revealed multiple cutaneous neurofibromata, café-au-lait spots and Lisch nodules, palpable thrill, and a bruit on the right side of the neck. Neurological examination revealed 0/5 power in the right upper limb and 4/5 power in the left upper limb, with brisk deep tendon reflexes in all four limbs.

Investigations
Doppler ultrasonography (Voluson 730, GE) of the right side of the neck demonstrated a high-flow arteriovenous fistula. Computed tomography (CT) (Siemens, Erlangen, Germany) and magnetic resonance imaging (MRI) (Siemens-Symphony, Erlangen, Germany) of cervical spine were done. CT scan showed erosion of the right pedicle [Figure 1A] of C4 vertebra. MRI demonstrated large areas of flow void extending from C4 to C6 with cord compression [Figure 1B]. Detailed digital subtraction angiography (DSA) (Siemens, Erlangen, Germany) of cerebral circulation was done subsequently. DSA demonstrated a high-flow vertebovertebral arteriovenous fistula (VV AVF) connecting the right vertebral artery (VA) at its origin to the vertebral venous...
plexus [Figure 2A]. There were multiple intersegmental collaterals bilaterally, supplied via right ascending cervical arteries [Figure 2B]. There were large venous pouches with drainage into the intraspinal extradural and paraspinal venous plexus. The anterior cerebral circulation was normal in all phases of the angiogram. There was no filling of the fistula from the anterior circulation. The vertebrobasilar circulation was normal otherwise, except the VV AVF.

**Treatment**

The patient was planned for embolization of the VV AVF with detachable coils under general anesthesia. During intubation, the patient was found to have short mento-thyroid distance. Hence, fiberoptic bronchoscopy-assisted intubation was done. A 6-Fr femoral short sheath was placed into the right common femoral artery. A 6-Fr envoy-guiding catheter (Codman Neurovascular, Raynham, Massachusetts, USA) was placed into the right subclavian artery. The fistula was cannulated with Echelon 14 microcatheter and Xpedion 14 wire (ev3 Endovascular, Inc., Plymouth, MN, USA) through the guiding catheter. The fistula was coiled by placing Axium 20 × 50 cm (ev3 Endovascular, Inc, Plymouth, MN, USA) as the first coil [Figure 3A]. The coil conformed well to the fistula, but there was no significant flow alteration. Ten more coils (Axium 20 × 40 mm, 18 × 40 mm, 16 × 30 mm, 16 × 30 mm, 14 × 40 mm, 14 × 30 mm, 12 × 30 mm, 10 × 20 mm, 8 × 16 mm, and 6 × 15 mm) were serially placed into the fistula under roadmap guidance. The flow in the fistula was slower compared to the pre-embolization state. However, further coils could not be pushed with repeated attempts, as the microcatheter came out of the fistula during the placement of the last coil. Right subclavian artery angiogram as well as left VA angiogram demonstrated residual fistula. Second session of embolization was planned.

**Stage II embolization**

The patient was observed for 3 months. At the end of 3 months, there was only a partial improvement in neurological status with significant residual deficits. Hence, an angiogram with possible stage II embolization was planned. The fistula was cannulated from the left VA through the vertebrobasilar junction into the right VA with the combination of Echelon 14 microcatheter with Xpedion 14 microwire [Figure 3B]. The fistula was coiled by serial placement of four coils (Axium 8 × 20 mm, 7 × 15 mm, 6 × 15 mm, and 5 × 12 mm) into the fistula and the adjacent VA. Check angiogram demonstrated residual filling of the fistula. subsequently it was decided to embolize the residual fistula with glue (n-butyl cyanoacrylate) and lipiodol from the ascending right cervical artery. For this, 66% glue was prepared by mixing appropriate amount of lipiodol. A total volume of 5 ml of glue was prepared and 3.5 ml of which was injected. The microcatheter was flushed with 5% dextrose. Under blank roadmap, the 66% glue was injected into the fistula in less than 45 sec. The check angiogram demonstrated complete exclusion of the fistula from the circulation [Figure 3C]. The final angiogram from the left VA and right subclavian artery demonstrated complete cure of the fistula with stable coil and glue mass [Figure 3D]. Patient was shifted to intensive care after the embolization and extubated after 6 h.
We did transarterial embolization of the fistula from producing more stable results with lesser complications. Onyx is a newer liquid embolic agent and allows a longer injection time with high volume of embolic materials, hence for successful occlusion of fistulas in some situations. A combination of coils and liquid embolics have been described for treatment of VV AVFs. A combination of coils and balloons or coils and glue/onyx can be used for successful occlusion of fistulas in some situations.

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We did transarterial embolization of the fistula from both the VAs with a combination of coils and n-butyl cyanoacrylate (glue). A complete occlusion of the fistula could be achieved without any complications and with a good clinical outcome.

**Conclusion**

VV AVF is a rare condition of abnormal connection between VAs and/or its branches and vertebral venous plexus. Endovascular treatments of VV AVF are safe and efficacious. High-flow VV AVF needs detailed angiographic study and proper planning for successful endovascular treatment.

**References**


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