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Spontaneous Vertebral Artery Arteriovenous Fistula with Neurofibromatosis Type I and Its Management with Covered Stent

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Abstract

Vertebro-vertebral fistulas (VVFs) are uncommon vascular pathology. It can be either primary (spontaneous) or secondary (iatrogenic or mechanical trauma). Spontaneous vertebral arteriovenous malformation is often associated with connective tissue disorders. Cases associated with neurofibromatosis type I (NF I) are even rarer. Management of VVF with covered stent is an emerging option for construction of vertebral artery. It not only preserves the flow of the parent artery but also has immediate exclusion of the fistula from the parent artery. A 30-year-old pregnant female patient presented with cervical bruit and left upper limb radiculopathy. She was a known case of NF I. Magnetic resonance imaging cervical spine revealed multiple flow voids compressing the cervical spinal cord and nerve roots. Digital subtraction angiography revealed a vertebral artery arteriovenous fistula. She underwent endovascular treatment in the form of a covered stent. Her clinical symptoms immediately improved. She was asymptomatic at the 1-year follow-up.

Keywords

- spontaneous vertebral artery arteriovenous fistula
- neurofibromatosis
- endovascular treatment
- covered stent
- arteriovenous malformation

Introduction

Vertebro-vertebral fistulas (VVFs) result when a direct shunt forms between the vertebral artery and the veins forming the vertebral venous plexus. These are uncommon vascular pathologies. They occur either spontaneously or due to trauma which can be mechanical or iatrogenic. Spontaneous VVFs are more often associated with congenital pathologies. Very few cases of VVF with neurofibromatosis type I (NF I) have been reported in the literature.[1] VVF can be located throughout the course of the cervical vertebral artery. However, spontaneous VVF are more commonly found at the level of the first cervical vertebra.[2] A covered stent is an emerging option for construction of vertebral artery, which is more efficacious than coils and embolic agents. It preserves the flow of the parent artery and has immediate exclusion of the fistula from the parent artery.[3] We report a case of spontaneous VVFs at the level of the fifth cervical vertebra in a young pregnant female with NF I, which was managed endovascularly with a covered stent.

Case Report

A 30-year-old female, 7 months primigravida (twin pregnancy) presented to our institute with complaints of neck swelling, bruit over the left side of the lower neck, and left upper limb radicular pain mainly over the outer aspect of the arm and forearm for 2 months. General examination

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revealed café-au-lait spots and dermal neurofibromas. Her neurological examination revealed power of grade IV at the left shoulder in abduction and left elbow in flexion, according to the Medical Research Council (MRC). Reflexes were exaggerated in both upper limbs and lower limbs. Babinski's sign was positive on both sides. Local examination revealed approximately 3 cm × 2 cm in size, soft, pulsatile swelling with palpable and audible bruit in the posterior triangle of the neck. Spine examination revealed thoracic scoliosis.

Magnetic resonance imaging (MRI) of the cervical spine revealed multiple, large flow voids located on the left side at the level of C4, C5, and C6 and also extending in to the spinal canal through neural foramen at the C4-C5 and C5-C6 level with the displacement of the spinal cord to the right. These MRI features were suggestive vertebral arteriovenous fistula between the left vertebra artery (V2 segment) and epidural cervical venous plexus (►**Fig. 1A,B**). Conservative management was opted considering her obstetric status. However, she underwent an elective caesarean section due to the intrauterine death of one of her fetuses. After 15 days post-surgery, she was shifted to the neurosurgery unit for further management.

She underwent computed tomography (CT) angiography of the neck vessels which revealed precise location of the fistula, its relation with neural foramen, and the diameter of vertebral foramens (►**Fig. 1C,D**). Her digital subtraction angiography (DSA) of brain and neck vessels revealed left vertebral (V2 segment) fistula at the C5-C6 level which was filling from both the vertebral arteries. It was also filling from the ascending cervical and transverse cervical branches of the left subclavian artery (►**Fig. 2A–C**). Venous drainage was noticed into the epidural cervical plexus, bilateral anterior jugular vein, bilateral external jugular vein, and bilateral internal jugular vein.

She underwent endovascular treatment in the form of covered stent under general anesthesia. She received the following loading dose of antiplatelets medications: 300 mg aspirin and 150 mg clopidogrel 6 hours prior to the

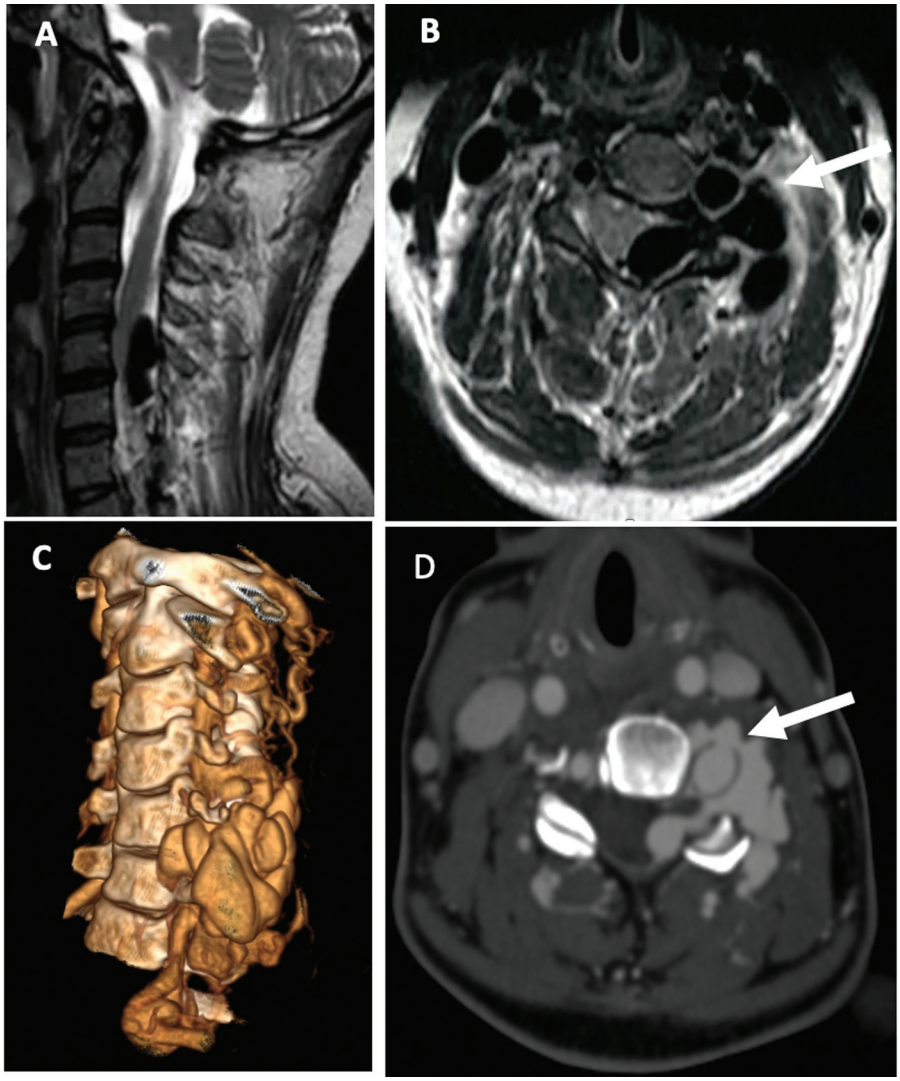

Fig. 1 Magnetic resonance imaging (MRI) scan of the cervical spine, T2-weighted sagittal image (**A**) showing multiple, large flow voids located at the level of C4, C5, and C6 mainly on the left side. MRI scan of the cervical spine, T2-weighted axial image (**B**) showing flow voids extending in to the spinal canal with displacement of the spinal cord to the right. Computed tomography (CT) angiography of the neck vessels oblique view (**C**) showing left vertebral artery terminating in to multiple fistulous pouches and axial view (**D**) showing site of fistulous communication.

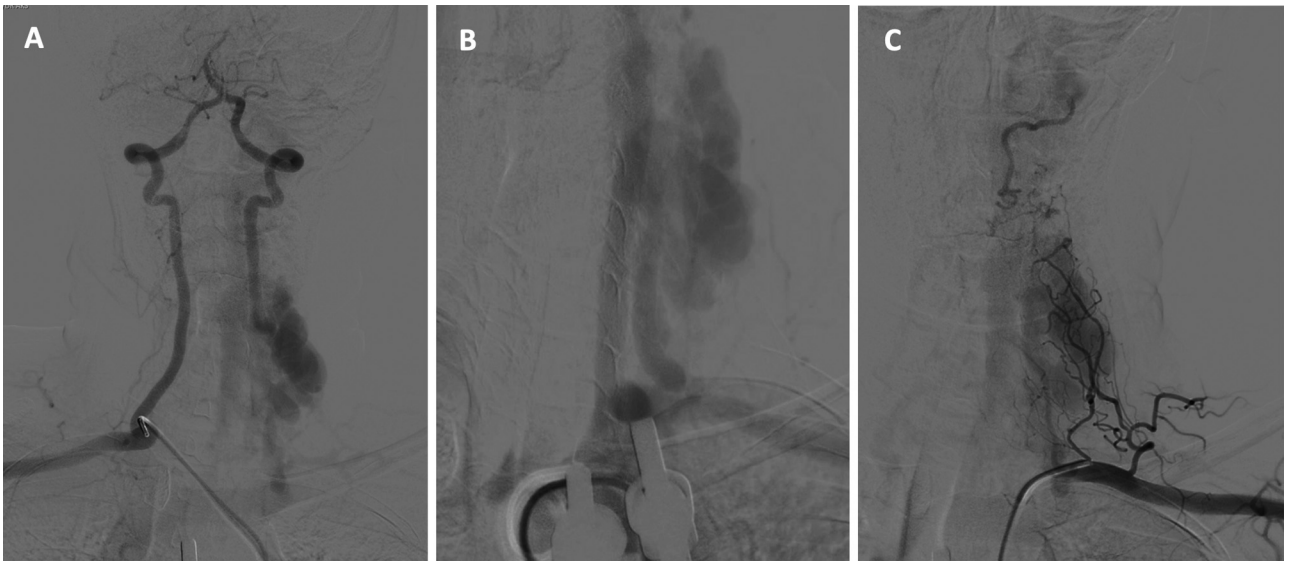

Fig. 2 Digital subtraction angiography (DSA) of brain and neck vessels showing left vertebral (V2 segment) fistula at the C5-C6 level which has forward flow from the left vertebral artery (**A**) and retrograde flow from the right vertebral artery (**B**). It was draining in to the left internal jugular vein. Fistula was also filling from the ascending and transverse cervical branches of the left subclavian artery (**C**).

procedure. An 8F arterial sheath was placed in the right femoral artery. About 5,000 units heparin was given intravenously. Left subclavian angiography was done. Under road map guidance, an 8F guiding (Envoy, J&J) catheter was positioned just above the origin of the left vertebral artery and the fistula site crossed with 035" curved hydrophilic guidewire (Terumo). A 6 mm × 40 mm size balloon-expanded covered stent (Bentley, InnoMed GmbH) was placed across the fistula site and deployed. Poststent deployment DSA run showed significant reduction of flow in the fistula (►**Fig. 3A,B**). Postprocedure, her neck swelling and left upper limb pain completely disappeared. The patient was discharged on day 5 postprocedure. Patient was treated with dual-antiplatelet medications (oral aspirin 150 mg and clopidogrel 75 mg) for 6 months and single antiplatelet (oral aspirin 150 mg) for life time. Power in the left upper limb improved to MRC grade V, 1 month postprocedure. At 1-year follow-up she was asymptomatic. Her follow-up DSA and CT angiogram revealed no evidence of the fistula (►**Fig. 4A–D**).

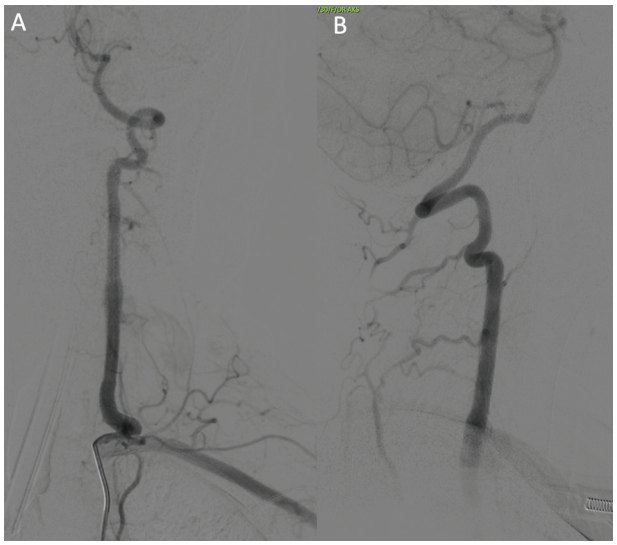

Fig. 3 Immediately after deployment of the covered stent, left vertebral injection run anteroposterior (AP) view (**A**) and lateral view (**B**) run showing near-complete obliteration of the fistula with patent left vertebral artery.

Discussion

VVF is a very uncommon pathology. Based on their formation, they can be categorized into spontaneous, traumatic, and iatrogenic in nature. Spontaneous VVFs are gradual in onset and progressive, whereas traumatic and iatrogenic VVFs are always acute. Spontaneous VVF is often associated with congenital lesions like NF I, fibromuscular dysplasia, or Ehler–Danlos syndrome. Till date, only 30 cases have been reported with VVF with NF I, including our case.[3] Etiologically, VVF in NF I may arise either as a result of mesodermal dysplasia or due to dysplastic smooth muscle and neurofibromatosis proliferation in arterial wall leading to aneurysm formation. Rupture of aneurysm into adjacent veins results in VVF.[4] Pregnancy may aggravate the symptoms related to the preexisting cerebrovascular disease mainly due to hemodynamic changes. Generally, cardiac output in pregnant women initially increases with gestation to peak at about gestational week 30, to levels approximately 50% greater than in nonpregnant controls.[5] Symptomatology depends on the site of the fistula and the flow within. Most of the VVFs are asymptomatic. However, VVFs can present with tinnitus, cervical bruit, headache, nausea and vomiting, vertebra basilar ischemia, or congestive cardiac failure.[6] Rarely, patient presents with symptoms of cervical radiculopathy due to direct compression of the nerve root by dilated draining veins or an aneurysm penetrating into the vertebral canal. These symptoms are most commonly seen in NF I.[4,7] MRI is the initial method of choice. MRI shows classical flow voids adjacent to the spinal cord that often causes compression of

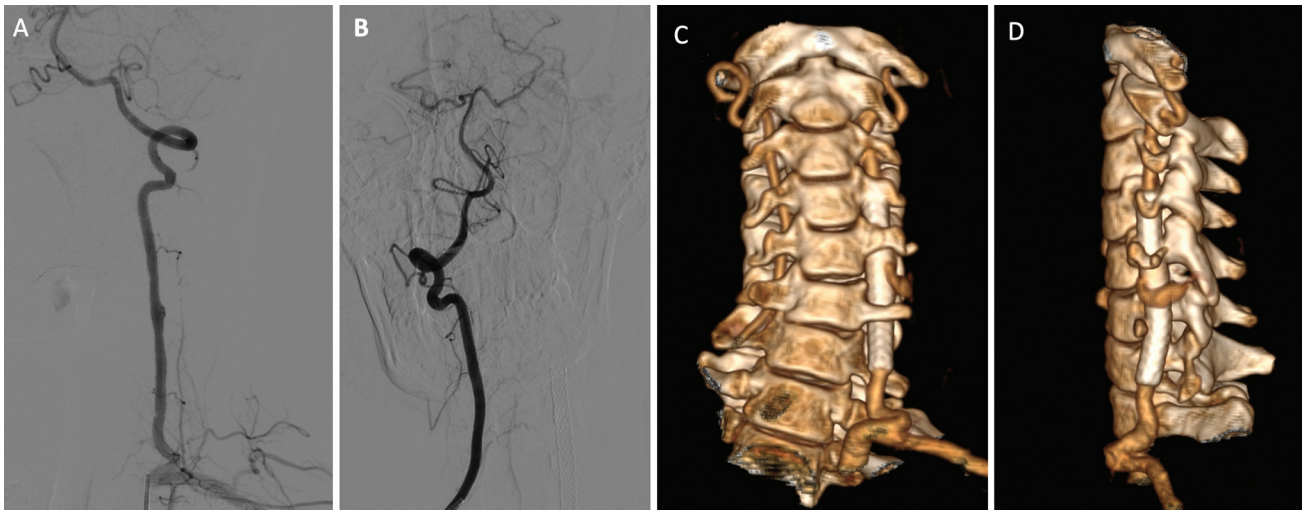

Fig. 4 Left vertebral injection run anteroposterior (AP) view (**A**) and right vertebral injection run lateral view (**B**) showing complete disappearance of the fistula. Computed tomography (CT) angiography of neck AP view (**C**) and lateral view (**D**) showing covered stent with nonfilling of the fistula.

the nerve root or spinal cord. DSA is the gold standard and provides information regarding angioarchitecture of the fistula including its location, size, flow along with feeding arteries and draining veins, as well as status of contralateral vertebral artery and its supply.[8]

Almost all VVFs require treatment as they may progress to cause symptoms later. Indications for treatment are based on symptoms and angioarchitecture of the fistula. The presence of neurological deficit due to spinal cord compression and radiculopathy due to nerve root compression are strong indications for the treatment. Presence of swelling and bruit in the neck often requires treatment as in our case. Treatment is also indicated if retrograde, intracranial, or spinal cord venous drainage is present.[9] Treatment options include microsurgery and endovascular procedure. Endovascular treatment is the therapeutic modality of choice and can be constructive or deconstructive. Constructive treatment includes occlusion of the fistula with detachable balloons and/or coils, covered stents, and flow diverters keeping the parent artery intact. Deconstructive treatment is usually considered in patients with adequate contralateral vertebral artery flow and involves occlusion of both the fistula and the parent artery.[10] Multiple reports are published suggesting complete occlusion of the fistula with covered stents. With the advent of endovascular therapy, microsurgery is rarely indicated. Surgery is usually considered when endovascular treatment fails and involves exposure of the fistulous tract and surgical ligation. Alternatively, surgical trapping of the vertebral artery can be considered if there is no other alternative and the opposite vertebral artery flow is intact.[11]

A self-expanding or balloon expanding covered stent is an emerging option for construction of vertebral artery, which is far more affordable and efficacious than coils. Very few case reports are published suggesting complete occlusion of the fistula with covered stents in cases of NF I.[3] Covered stents are either self-expandable or balloon-expandable. A balloon-expandable stent is expected to be more attachable to blood vessels, but there is a risk of injury such as dissection of implanted vessels. Also, balloon-expandable delivery systems lack flexibility, a self-expandable stent is more appropriate if the target vessel is movable. Advantages of a covered stent include preservation of parent artery and risk of mass effect on adjacent structures by use of embolic materials and coils can be avoided.[12] Other advantages include covered stents have a good flexibility and efficacy in the lower vertebral region (V1-V2 segment). Covered stents also work well with the regeneration of vascular intima. Use of covered stents in the upper vertebral region (V3-V4 segment) is usually avoided due to extreme tortuosity of the artery and chances of kinking, fracture, or migration of the stent in this region. Other minor risks include chances of endoleak and use of antiplatelet agents for the whole life.[13] In our case, we have used a covered stent in the V2 segment of the vertebral artery under antiplatelet cover.

Conclusion

VVF is a very uncommon pathology. Based on their formation, they can be categorized into spontaneous, traumatic, and iatrogenic. Spontaneous VVF is often associated with congenital lesions, most commonly NF I. Endovascular treatment is the therapeutic modality of choice. The use of covered stent is a safe and durable treatment option for the preservation of the parent artery. It is more efficacious and affordable than coils and other occluding agents.

Conflict of Interest
None declared.

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