A VVF is a rare type of spinal arteriovenous fistula that occurs in the cervical region. It is an abnormal communication between the VA and its branches with the adjacent venous system. Most VVFs are post-traumatic or iatrogenic in origin; spontaneous fistulas are rare and are usually seen in vascular dysplastic conditions like NF. Various endovascular treatment options such as balloon embolization, coiling, and covered stent placement are available for successful management of these fistulas. Here we report the combined transarterial and percutaneous coiling of a VVF in a patient with NF1. This case is unique and challenging because of previous failed surgical attempts.

**Case Report**

**History and Examination.** This 29-year-old woman with a known case of NF1 presented with a 3-month history of noting a humming sound over the left side of her neck, radicular pain in her left upper limb, paraesthesia, and progressive weakness of both upper and lower limbs, more so on left side. An MR imaging study showed an enlarged cervical epidural venous sac extending from C-3 to C-6 that compressed the spinal cord and caused cord hyperintensity on T2-weighted images (Fig. 1).

Digital substraction angiography showed a high-flow 12-mm fistula with a large distal venous sac (32 × 36 mm) connecting the left VA and an emissary radicular vein at the level of the C5–6 intervertebral foramen. The fistula drained into dilated cervical epidural venous sacs from C-3 to C-6 and subsequently to extravertebral veins and the internal jugular vein. Surgical trapping of the left VA above C-2 and below the origin was attempted without success. Subsequently, the VA was ligated at its origin. After the surgical ligation, CT angiography followed by DS angiography revealed tight narrowing of the left VA at its origin and occlusion at the level of C-2. The proximal left VA was dilated and tortuous with absent antegrade flow beyond the fistula. Supply to the fistula was noted via the V₃ segment through collateral vessels (Fig. 2). Because of the persistence of the fistula and consequent neurological deterioration, the patient was referred to our institution.
On arrival, the patient was bedridden. A general examination revealed cutaneous neurofibromata, café-au-lait spots, palpable thrill, and a prominent bruit over the left side of her neck. Neurological examination showed grade 0–1/5 power in both upper limbs and 3/5 in both lower limbs with increased tone in the right upper and both lower limbs. Additional DS angiography studies performed at our institution confirmed the persistence of a high-flow left VVF as well as multiple intersegmental collaterals ascending bilaterally over the cervical, deep cervical, occipital, and right VA.

Operation. The initial plan was to occlude the V2 segment of the VA in the fistulous segment starting from distal to proximal aspect. A 7 Fr introducer sheath (Terumo Corp.) was placed in the right common femoral artery route, and the left VA was catheterized with a 4 Fr VA glide catheter (Terumo Corp.). Attempts at catheterizing the VA at its postfistulous segment were unsuccessful. Hence, the Excelsior 18 microcatheter (Boston Scientific) over a Transcend 0.010-in microguidewire (Boston Scientific) was placed into the postfistulous venous sac, and coiling was attempted with GDC-18 standard coils (15 mm × 30 cm; Target, Boston Scientific). However, owing to the high-flow fistula and large size of the venous sac, the coil migrated distally into the dilated intraspinal epidural venous sac without staying in the extraspinal sac. The ligated origin of left VA was dilated, and a 7 Fr guiding catheter (Launcher, Medtronic, Inc.) was negotiated into the proximal left VA, which led to an immediate reduction of the flow across the fistula. Two microcatheters (Excelsior 18) were introduced into the postfistulous venous sac, and 2 GDC-18 standard coils (20 mm × 30 cm) were deployed simultaneously. This was followed by placement of GDC-18 standard coils of varying sizes and 5 stainless steel coils (size 52-15-15, Cook, Inc.) into the same extraspinal postfistulous venous sac. This allowed us to pack the venous sac and the proximal left VA. Angiography showed total obliteration of the flow in fistula from proximal left VA; however, we noted some retrograde filling of the fistula (Fig. 3a–c). Because further coil deployment was not possible transarterially, we decided to obliterate the left V2 segment through a percutaneous approach in subsequent sitting.

Four days later a direct puncture of the left V2 segment was done under fluoroscopic guidance with a 16-gauge direct puncture needle (Bio-Med, Inc.). However, the 4 Fr sheath (Terumo Corp.) could not be placed in the VA. The postfistulous venous sac was directly punctured with a 16-gauge Seldinger needle (Eastern Medikit Ltd., India), and the cannula was left in situ. An Excelsior-18 microcatheter was passed through the cannula, and 5 GDC-18 standard coils of various sizes were deployed in the sac. The last coil extended into the V2 segment. An angiogram showed total obliteration of the fistula and nonfilling of the cervical epidural venous plexus (Fig. 3d).
Coiling of a spontaneous VVF associated with NF1

Fig. 3. Angiograms. a: Image obtained before coiling of the left subclavian artery, showing the high-flow fistula with a large postfistulous venous sac. Shunting of blood to the epidural venous sac can be seen through the C5–6 neural foramina (arrows). b: Postcoiling left VA injection demonstrating absent antegrade filling of the fistula; however, retrograde filling of the fistula can be seen through the cervical collaterals (arrowheads) because of leakage of contrast material into the subclavian artery. c: Right VA image obtained after coiling, revealing retrograde filling of the left V2 segment (arrowheads) and the fistula through intersegmental collaterals (arrow). d: Right VA image following percutaneous coiling demonstrating nonfilling of the fistula. Note the percutaneously placed cannula (white arrow) with microcatheter (black arrow) in the coil mass.

Postoperative Course. The peri- and postprocedural periods were uneventful. Three days after the second procedure, the patient showed signs of improvement with an increase in upper limb power bilaterally. She was discharged from the hospital and underwent proper physiotherapy training. At the 3-month follow-up, the patient showed significant clinical improvement and was ambulatory. At 1 year of follow-up, the patient had no neurological deficit and had normal power in both upper and lower limbs. Doppler ultrasonography showed absence of flow in the fistula.

Discussion

Vertebrovertebral fistula is a term used to describe various types of fistulas that involve vertebral arteries and veins. It is classified under the group of parachordal arteriovenous fistula. Most of these (68%) are of traumatic or iatrogenic in origin, and 32% are spontaneous. Spontaneous variants are less common and are associated with vascular dysplastic conditions such as neurofibromatosis, fibromuscular dysplasia, and Ehlers-Danlos syndrome. The underlying cause in these cases is arterial, and the fistula is microtraumatic. There are a limited number of case reports of a spontaneous VVF associated with NF1. The majority of patients are female. They commonly present with symptoms of NF1 (100%), radiculomyelopathy (78%), and bruit over the neck (50%). Other less common symptoms include tinnitus (10%), cranial neuropathy (3%), or a pulsatile neck mass (3%). The symptoms noted in the present patient were due to compression of the spinal cord by the pulsatile neck mass and the dilated ectatic epidural venous sac.

Vertebrovertebral fistulas can be classified either as segmental and intersegmental types or as upper and lower cervical groups. In the segmental type, the fistula involves the VA branch, whereas in the intersegmental type, the VA itself is involved. The upper cervical group usually seen in the pediatric population includes a fistula that has developed at the C1–2 level from the proatlantal system. Lower cervical fistulas involve cervical arteries and can be seen in adults with underlying dysplastic vessel wall disease. These fistulas occur at the level of the vertebral canal and drain into the epidural venous plexus, which can lead to formation of enlarged epidural venous lakes, causing spinal cord compression. Our case can be classified as an intersegmental type of lower cervical VVF. Also, if the lesion is categorized according to the flow pattern in the VVF as described elsewhere, the fistula in the present case could be stated as having a Type II pattern.

It goes without saying that to plan a definitive management of such pathological entities, an angiogram is necessary for categorizing the fistula as one of the aforementioned types. There could be a surgical option or an endovascular approach for its management. Surgical treatment consists of resection or trapping of the fistula. It is difficult and dangerous due to the proximity of surrounding neurovascular structures, which often renders a surgical approach impossible. However, at surgery the fistulous communication should be correctly localized and excluded to avoid blockage of the endovascular route for future interventions in case of recurrence, as happened in our case. Recurrence is generally caused by recruitment of intersegmental collaterals from other cervical arteries.

Treatment has evolved considerably in recent years, and endovascular occlusion of fistulas is now the treatment of choice. Surgery alone or in conjunction with an endovascular procedure may be useful for treating complex lesions.

Various endovascular techniques are available for the management of VVFs. Commonly described techniques include transarterial use of detachable balloons, transarterial placement of detachable coils at the fistulous site with or without trapping of the parent vessel, and placement of a covered stent in the feeding artery. Other than these aforementioned transarterial approaches, adjunctive transvenous and direct percutaneous embolization has been used to close VVFs. One should opt for the technique that could match the size and hemodynamic nature of the fistula, resulting in its complete obliteration.

Methods such as placement of detachable balloons in cases similar to the present one are dangerous because of the chance of distal migration into the venous side, in view of the large size and high-flow nature of the fistula. Similarly, postsurgical complex anatomy rendered the
placement of covered stents across the fistula impossible. Therefore coiling of the extraspinal fistulous venous sac and the parent VA with GDC coils was preferred. Use of detachable coils is safe and easy, and they allow better control, particularly in large high-flow fistulas.\(^6^,\(^1^,\(^3\))

It is necessary to preserve the patency of the parent artery in young patients or in those who cannot tolerate vessel closure;\(^6^,\(^1^,\(^3\)) however, in VVF with large high-flow fistulas, maintaining the patency of the parent artery is difficult.\(^6^,\(^1^,\(^3\)) Occlusion can be done either through an antegrade or retrograde approach using coils and/or balloons.\(^6^,\(^1^,\(^3\)) Difficult catheterization of the distal artery through an ipsilateral route and blocked arterial access from the contralateral side posed difficulty in coiling of the distal artery; therefore, we decided to coil the postfistulous venous sac and the proximal VA.

We successfully placed the GDC coils with the simultaneous use of 2 microcatheters, which helped us to form a basket for further coil deployment. This method has been well described in managing wide neck large aneurysms.\(^8^) In our opinion, it is also helpful in such large high-flow fistulas, because the basket prevents distal coil migration. Furthermore, the use of high-volume Cook stainless steel embolization coils reduced the number of GDC coils used, which reduced the procedural cost. We believe that high-volume coils can be useful in such large fistulous sacs because they do not migrate and provide a good filling volume, although in the present case, the delivery was difficult and cumbersome. In view of the difficulty in placing coils further and retrograde filling of the fistula, we used the percutaneous approach for coil placement and successfully closed the fistula. To our knowledge, this kind of combined transarterial and percutaneous coiling has not been reported in the literature. However, in this unique and challenging case complicated by previous surgery, it helped us to achieve complete closure of the fistula. This experience may be helpful to others in managing such difficult cases.

The present case points out the challenges encountered in treatment of VVF complicated by previous failed surgery. In cases such as this, combined endovascular and direct percutaneous coiling is possible and may help in treating such complex fistulas.

**Disclaimer**

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

**References**