Neurofibromatosis type 1 (NF-1) is one of the most common autosomal-dominant genetic disorders with an incidence of approximately 1:3,500. Vascular abnormalities associated with NF-1 include stenotic or occlusive diseases, aneurysms, arteriovenous malformations and, rarely, arteriovenous fistula (AVF). Association of arteriovenous fistula with NF-1 is rare with a few cases of vertebral AVF having been reported (1–3). Endovascular occlusion of the vertebral fistula offers a minimally-invasive treatment option with an acceptable number of possible complication and results. We report a case of spontaneous vertebral AVF with dilated epidural venous plexuses causing cervical cord compression resulting in quadriparesis. The patient was successfully treated by embolization of the fistula with coils and a balloon.

CASE REPORT

A 42-year-old woman presented with complaints of neck pain of four months’ duration, gradual onset of retroauricular bruit, and weakness of all four limbs during the past two months. She could not remember any traumatic history. On examination, she had multiple, soft, cutaneous nodular swellings and brownish spots all over her body, both suggestive of neurofibromatosis. Plain films of the cervical spine showed increased, lobulated suboccipital and paravertebral soft tissue density, although the cervical bony structures appeared normal. MRI of the brain revealed multiple, large, flow voids in both the epidural space and the paravertebral region of the upper cervical spine, both suggestive of dilated epidural and paravertebral vascular channels (Fig. 1A). These dilated vascular sacs were noted to be compressing the cervical cord, most significantly at the C1-C5 levels. The MIP (maximum intensity projection) image of CT angiography revealed vertebral AVF with dilated, epidural venous plexuses compressing the cervical cord (Fig. 1B). A diagnosis was made of vertebral AVF in NF-1 with a dilated, epidural venous sac causing compression of the cord. These findings were confirmed on digital subtraction angiography. A single, high-flow
AVF at the C2 level with a direct shunt from the right vertebral artery to the epidural veins, was verified. Large, ectatic venous pouches drained further into the dural and paravertebral venous plexuses (especially on the right side) (Fig. 1C, D). The fistula was filled from both internal carotid arteries and from the left vertebral

Fig. 1. A. A 42-year-old female presented with quadriparesthesia and retroauricular bruit. The patient exhibited subcutaneous plexiform neurofibromas. Cervical MR images show dark signal intensities of the signal voids caused by an engorged epidural vein (thin arrow) compressing the spinal cord. Note the right subcutaneous neurofibroma (thick arrow). B. Sagittal reformatted MIP image of the CT angiogram showing a dilated epidural venous sac from C1 to C5 (arrows) and compressing the spinal cord. C. Left vertebral angiography shows the steal phenomenon to the fistula (arrow) without filling the bilateral posterior cerebral artery. D. Right vertebral angiography reveals an enlarged VA and a high-flow, arteriovenous fistula at the C1–2 junction. A fistulous hole is directed medially and drains into the engorged epidural vein and the paravertebral venous channel. E. The coil basket was positioned distal to the fistula via the contralateral left vertebral artery. F. Post-embolization oblique projection view showing the balloon (arrow) on the right side. G. Post treatment right vertebral angiography shows cessation of flow in the caudal vertebral artery. H. Post embolization left vertebral angiography shows complete occlusion of AVF and restoration of antegrade left VA flow.
artery through collaterals of the Circle of Willis (not shown). External carotid angiograms bilaterally showed no remarkable arterial supply to reconstitute the fistula. Different treatment options were considered, and embolization was determined to be the best treatment option. Informed consent was obtained from the patient and her family. Entrapping of the right vertebral AVF was planned, and embolization was then performed under light sedative anesthesia two weeks later. Bilateral femoral access was secured. A 6-F sheath was placed in the left femoral artery, and a 6-F Envoy catheter (Cordis Neurovascular, Miami Lake, FL, U.S.A.) was inserted into the left proximal vertebral artery. A Prowler plus microcatheter (Cordis Neurovascular, Miami Lake, FL, U.S.A.) was navigated into the distal right vertebral artery. The catheter tip was positioned in the right vertebral artery just distal to the fistulous hole. The distal segment of the venous pouch of the right vertebral artery was occluded using DCS-18, mechanically detachable platinum coils (William Cook Europe, Bjaeverskov, Denmark) as well as push-type fibered coils (Fig. 1E). Complete occlusion of the distal fistulous segment of the right vertebral artery was accomplished after a total six coils were inserted. An 8-F Guider (Boston Scientific, Fremont, CA, U.S.A.) was introduced through the right femoral sheath and placed into the origin of the right vertebral artery. The proximal segment of the large venous pouch in the right vertebral artery was embolized using a detachable balloon GVB 19 (Nycomed Amersham, Paris, France) (Fig. 1F). The post-embolization angiogram showed complete occlusion of the right-sided vertebral AVF (Fig. 1G) and good antergrade flow in the left vertebral artery (Fig. 1H). At six months post-embolization, the patient had improved symptoms and was ambulant with support.

**DISCUSSION**

Vertebral AVFs are abnormal shunts between the extracranial vertebral artery (or one of its muscular or radicular branches) and a neighboring vein. These rare lesions can be traumatic or spontaneous in origin. Various connective tissue diseases including neurofibromatosis, Marfan’s syndrome, and fibromuscular dysplasia, are known to be predisposing factors (1–3). Cervical fistulas have a unique predilection to develop in patients with NF-1. Unlike the dural type-1 fistula, 97% of spontaneous spinal AVFs associated with NF-1 originated from the vertebral artery in the cervical spine, none was found in the thoracic or sacral spine, and only one occurred as a lumbar lesion (1).

In the evolution of an AVF in NF-1, dysplastic smooth muscle or neurofibromatosis proliferation in the wall leads to vasculopathy, aneurysm formation, leakage, and ultimately rupture into adjacent veins. If there is a fistulous connection between the vertebral artery and vein, pressure changes lead to dilatation of theseplexuses. The engorged, pulsating venous sacs in the epidural space can exert a compressive force on the spinal cord and thus contribute to cord compromise and lead to radiculopathy (4).

With the development of intravascular embolization techniques, occlusion with detachable balloons and coils is considered to be a safe and reliable method for treating AVF, and with significant reduction in morbidity and mortality that could be caused by surgical therapy (5–8). Obvious advantages of endovascular therapy include the ability to perform the procedure under local anesthesia, thus allowing continuous monitoring of the patient’s condition, especially if parent vessel occlusion is necessary.

The goal in managing epidural AVF in NF-1 is complete occlusion of the fistula, and preservation of the patency of the vertebral artery. Occlusion can be done using either an antegrade or a retrograde approach using coils and/or a balloon. To reduce the high flow into the fistulous hole and to precisely delineate the fistula site, we first placed the microcatheter distal to the fistula via the left vertebral artery using a cross-over technique and then occluded the distal fistulous channel. We successfully placed the DCS coils, which helped to form a basket for further coil deployment.

This is very helpful with such a large, high-flow fistula, as the basket prevents coil migration into the fistula. Furthermore, use of push-type, fibered coils reduced the number of detachable coils, which also reduced the cost of procedure. Considering the proximity of the parent artery to fistulous venous pouch, we occluded the proximal segment of the large, venous pouch of the right vertebral artery using a balloon and then successfully closed the fistula. Occlusion of the venous pouch with balloons or coils would also obliterate the fistula, although this would require packing of the large venous sac at the intervertebral foramen with balloons or coils, which might tighten the foramen and compress the nerve root.

Accessibility is an important consideration in such a neuro-endovascular procedure. The balloon needs a large guiding catheter. In this case, the proximal portion of the right VA was larger than usual because of the
large arteriovenous shunt through the fistula, which allowed us to use a detachable balloon.

Since its introduction in the late 1970’s, detachable balloon embolization has been widely used as a cost-effective endovascular treatment. However, the balloon configuration may be inadequate to fill and occlude the entire fistula pouch, thus requiring additional embolization using a coil or N-butyl 2-cyanoacrylate (7). Moreover, detachable balloons may deflate or migrate into the venous side, due to the large size and high-flow nature of a fistula. Therefore, coiling of the extraspinal fistulous venous sac and parent vertebral artery using a detachable coil is preferable. Use of a detachable coil is safe and easy, and allows better control, particularly in large high-flow fistulas (8, 9).

A reported potential complication with endovascular treatment of a vertebral AVF is perfusion pressure breakthrough resulting in cerebral hemorrhage. On abrupt closure of the fistula, normal perfusion pressure breakthrough can result in neurological dysfunction and/or hemorrhage (10). Because of this concern, we maintained strict blood pressure control during the patient’s admission to the intensive care unit. The illustrated case points out the complexity of clinical disorders encountered with NF-1. With diagnostic imaging, a complementary CT angiography, cross-sectional MRI in addition to selective angiography could depict the anatomical details.

In conclusion, this is a rare case of vertebral AVF in NF-1, with successful endovascular treatment that clearly demonstrates the advantages of endovascular techniques. Endovascular occlusion of vertebral AVFs is a safe, reliable, cost-effective, and reproducible method which is now the primary treatment for this condition. It can be performed with close neurologic monitoring in conscious patients, and with minimal morbidity and no mortality.

References
제1형 신경섬유종 환자에서의 자발성 척추동맥 동정맥루의 혈관내 치료 : 증례 보고

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자발성 척추동맥 동정맥루는 제 1형 신경섬유종 환자에서 드물게 동반되는 혈관 병변이며, 배출되는 경막외 동맥의 확장으로 인해 다양한 신경학적 증상이 유발된다. 저자들은 신경근척수병증을 보인 42세 신경섬유종 환자에서 코일과 풍선을 이용하여 동정맥루를 성공적으로 치료한 사례를 보고하고자 한다.

Key Words : Neurofibromatosis; Vertebral arteriovenous fistula; Embolization