Targeted Embolization of Vertebral Arteriovenous Fistula Associated with Type 1 Neurofibromatosis: Case Report

Yoshihiko Watanabe, Hidenori Miyake, and Hideki Ogata

Objective: Vertebral arteriovenous fistula (AVF) is rare in type 1 neurofibromatosis (NF1). We report a case of NF1 complicated by vertebral AVF, for which transarterial targeted embolization of the fistula resulted in a favorable outcome.

Case Presentation: A 44-year-old man with NF1 presented with cervical radiculopathy of the right fifth nerve root due to vertebral AVF. The fistula was completely occluded by transarterial targeted coil embolization of venous pouch of the fistula, and his neurological deficits got better. After a year follow-up, the fistula was completely occluded, and the patient's radiculopathy recovered almost completely.

Conclusion: Targeted embolization via artery can be the first-line treatment method for vertebral AVF associated with NF1.

Keywords ▶ type 1 neurofibromatosis, arteriovenous fistula, radiculopathy, targeted embolization

Introduction

Type 1 neurofibromatosis (NF1) is an autosomal dominant hereditary disease that presents with a diverse range of symptoms. Of these, while vascular lesions are not uncommonly observed, vertebral arteriovenous fistula (AVF) is rare in NF1.1–3) A few cases have been cured by means of endovascular treatment. Detachable balloons or a number of coils have been used in these cases. Here, we report a case of NF1 complicated by vertebral AVF, for which transarterial targeted coil embolization of the fistula resulted in a favorable outcome.

Case Presentation

The patient was a 44-year-old man with weakness and discomfort from the right shoulder to the right upper arm. Past medical history was significant for NF1 and hyperlipidemia. The patient’s family history revealed that his mother had NF1. Regarding the present complaint, the patient had noticed a feeling of discomfort and weakness from the right shoulder to the right upper arm. His local doctor had referred him to our Department of Orthopedic Surgery, where he was examined. Neuroradiological examination suggested a suspected AVF in the cervical vertebrae, and the patient was therefore referred to our department.

On admission, the patient showed symptoms of cervical radiculopathy of the right fifth nerve root with a score of 3/5 on manual muscle testing of the right deltoid and right biceps brachii muscles. There were no spinal cord signs and abnormal tendon reflexes in any of the limbs.

MRI of the cervical vertebrae revealed a vessel that was dilated in the right anterior direction within the spinal canal from the level of the first to fifth cervical vertebrae, resulting in extradural compression of the spinal cord, in which no abnormal signals were identified (Figs. 1A and 1C). On 3D-CTA, we observed an AVF originating from the right vertebral artery, which was drained through intervertebral foramen into a dilated extradural vein within the spinal canal (Figs. 1B and 1D). On angiography, dilation of the right vertebral artery, an AVF, and a venous pouch at the level of C4/5 were observed (Fig. 2A). Angiograms of the left vertebral and right deep cervical artery also showed findings of an AVF at the same site, indicating a single fistula (Fig. 2B).

On endovascular treatment, although the contralateral vertebral artery was patent, the aim was to preserve the parent artery because the patient was relatively young. We also took into consideration the fact that we had to avoid
the risk of distal embolism due to incomplete parent artery occlusion. Therefore, we planned to perform transarterial targeted embolization by tightly packing the venous pouch. Treatment was performed under local anesthesia after placing the patient under mild sedation with midazolam. We punctured the right femoral artery and placed a guiding catheter with balloon at the distal tip (8-Fr OPTIMO; Tokai Medical, Inc., Aichi, Japan) into the right vertebral artery. The patient was heparinized to an activated clotting time of approximately 250–300 seconds. We placed the balloon catheter, Scepter XC (Terumo, Tokyo, Japan) distal to the right vertebral artery shunt and dilated the OPTIMO and Scepter balloons so that the fistula could be trapped. Next, we passed the microcatheter, Excelsior SL-10 (Stryker, Kalamazoo, MI, USA) into the fistula via the vertebral artery and guided it into the venous side within the spinal canal (Fig. 2C). To prevent overinflation, the diameter of inflated balloons was determined with the superimposed image of the right vertebral artery. While the OPTIMO and Scepter XC balloons were kept being dilated, coil embolization of the venous pouch was initiated. The entire venous pouch was framed using the microcoil, CASHMERE, measuring 8 mm × 20 cm (Codman Neuro, Johnson & Johnson, MA, USA). Subsequently, tight internal packing was performed using the flexible DELTAPLUSH microcoil (Codman Neuro) as tightly as possible. As we kept the view angle by which the right vertebral artery and venous pouch were clearly separated, tight packing was possible without balloon assistance. A total of 10 coils were used for embolization. To prevent coil migration in such case of high flow AVF, we first deflated the Scepter balloon, and deflated the OPTIMO balloon as slowly as possible.
Targeted Embolization of Vertebral AVF

We have seen numerous reports stating that the first choice of treatment for AVF associated with NF1 is considered to be endovascular treatment, and direct surgery is rarely required.1,6,7) But in these reports of endovascular treatment, most of the cases were treated with detachable balloons which occluded parent arteries, and no case was treated by targeted embolization which occluded only venous pouch.

In the present case, we considered the possibility of postoperative symptomatic exacerbation due to propagation of venous thrombosis to normal venous circulation.8) We therefore administered anticoagulation therapy for 1 week postoperatively. But anticoagulation might be unnecessary because the patient had no pial reflux into veins of spinal cord in the present case.

In the present case, because the orifice of the fistula was relatively narrow and the distance between intervertebral foramen and venous plexus was relatively long, we were able to plan and perform targeted embolization. But in such case as of a broad fistula or of short distance between intervertebral foramen and venous plexus, occlusion of parent artery or application of stent graft might be needed.

When we selected targeted embolization in the present case, we could not rule out the possibility that the radiculopathy would not improve due to compression with the coil mass. If we had occluded the parent vessel, this risk may have been reduced. We performed targeted embolization in the present case, and as a result, the symptoms gradually improved postoperatively. After 1 year, the patient had almost completely recovered. Plain X-ray images show the coil mass occupying only the upper half of right intervertebral foramen of C4/5 (D).

We planned to perform occlusion of the parent artery, if occlusion could not be achieved with the above method. But after deflation of the balloons, angiography of the vertebral artery showed complete obliteration of the AVF and preservation of the vertebral artery (Fig. 2D).

Postoperatively, anticoagulation therapy was employed using heparin and warfarin for 1 week due to concerns regarding propagation of venous thrombosis causing disturbance of normal venous circulation. Fortunately, the patient’s subsequent clinical course progressed without any worsening of neurological symptoms and discharged home 2 weeks later. MRI after 1 year showed complete occlusion of the AVF (Figs. 3A–3C). In terms of muscle strength, the right deltoid has completely recovered, and although slight muscle weakness remains in the right biceps brachii, there is a tendency toward improvement.

Discussion

NF1 is an autosomal dominant hereditary disease with a prevalence of 1:3500. It presents with a variety of clinical findings and is commonly accompanied by vascular lesions. Vascular lesions typically include arterial occlusion, intracranial vascular hypoplasia, moyamoya vessels, and arterial aneurysms.1) AVFs associated with NF1 are rare, and to date, there have been only 40 case reports,2,3) of which a few have been cured by means of endovascular treatment. The cause of AVF is usually believed to be an acquired condition caused by vascular fragility associated with NF1,4) but some believe that it is due to congenital abnormalities of arteriovenous communication.5) In the present case, MRA of the patient’s mother, who had NF1, revealed an unruptured vertebral artery aneurysm at the level of cervical vertebra. Therefore, we presume that the former mechanism is involved in AVF formation.

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Fig. 3 After 1 year, MRI T2-weighted image sagittal view (A) and axial view at the level of C4/5 (B) showed normalized spinal cord. The coil mass was pointed by arrow. MRA showed complete obliteration of the fistula and preservation of the right vertebral artery (C). X-ray photography showed the coil mass occupying only the upper half of right intervertebral foramen of C4/5 (D).
elimination of pulsatile stimulation to the cranial nerve is associated with improvement in symptoms.9) We presume that a similar mechanism was involved in the present case. We therefore believe that targeted embolization could become a first-line treatment, especially in cases with hypoplasia of the contralateral vertebral artery.

**Conclusion**

We reported a case of vertebral AVF associated with NF1. Identification of the fistula facilitated targeted embolization. We were also able to preserve the parent vessel and observed improvement in the radiculopathy. Targeted embolization via artery can be the first-line treatment method for vertebral AVF associated with NF1.

**Disclosure Statement**

The authors declare that they have no conflict of interest.

**References**


