Neurofibromatosis type 1 is well known to be one of most common autosomal dominant genetic disorders and this inherited disease is associated with various neoplasms, skeletal and vascular abnormalities (1). The vascular lesions affecting the head and neck of patients with neurofibromatosis type 1 (NF-1) has been reported as occlusive or stenotic form, moyamoya vessel or aneurysm. However, spontaneous vertebral arteriovenous fistula (AVF) associated with NF-1 is relative rare but characteristic manifestation (2). As sited adjacent to the cervical spinal canal, a vertebral AVF may drained via the epidural venous plexus, which may dilated and result in compression of the adjacent spinal cord and nerve roots. Endovascular occlusion of the fistulous hole offers a acceptable results and complication rate. We present a case of NF-1 patient with spontaneous vertebral AVF causing cervical cord compression. The patient was treated with endovascular coiling successfully.

**CASE REPORT**

A 44-year-old male presented with numbness in his both hands and radiating pain of right posterior neck and shoulder for two months. He had history of two surgical resections of soft tissue mass about 15 to 20 years ago. There was no history of trauma he could remember. The feature of this patient is characteristics of neurofibromatosis, including caf? au lait spots, multiple subcutaneous neurofibromas. Plain film of cervical spine showed increased lobulated paravertebral soft tissue density but cervical bony structures are normal-looking. The MR images of the cervical spine revealed large signal void in the right epidural and paravertebral area. These engorged epidural vein compressed the spinal cord at C2, 3 and upper aspect of C4 level most significantly (Fig. 1A & 1G). However, signal change of the cord at this level is not apparent. A right vertebral angiography depicted a high flow arteriovenous fistula at the medial aspect of C1-2 junction level with very tortuous enlarged proximal vertebral artery (Fig. 1B). The large,
engorged epidural venous sac drained to paravertebral venous plexus and internal jugular channel. In addition to the direct antegrade flow from the right vertebral artery, this fistula was filling from ipsilateral internal carotid artery, occipital artery and contralateral vertebral artery (Fig. 1C).

Informed consent was obtained from the patient and his family. Entrapping of the right vertebral AVF was planned and coil embolization was performed under intravenous anesthesia five days later. A temporary balloon occlusion test was not taken because preserved left vertebral artery is estimated to be large enough to supply the post territory. Bilateral femoral access was done. A 6-F sheath was placed in the right femoral artery, and 6-F guiding catheter (Envoy®, Cordis Endovascular, Miami Lake, FL) was forward into the proximal vertebral artery. Another 4-F diagnostic catheter was inserted through the opposite left femoral

![Image A](image1.png)

![Image B](image2.png)

![Image C](image3.png)

![Image D](image4.png)

**Fig. 1.** A forty-four year old male presented with tingling sensation of both hands and radicular pains. The patient shows cafe au lait spots and subcutaneous plexiform neurofibromas. Cervical MR images (A & G) show dark signal intensities of signal voids caused by engorged epidural vein (black arrow) compressing the spinal cord. Note the right subcutaneous neurofibromas (white arrows). Right vertebral angiography (B) reveals a tortuous and enlarged VA and high flow arteriovenous fistula at the C1-2 junction. Fistulous hole directed medially and drains into the engorged epidural vein and paravertebral venous channel. Left vertebral angiography (C) shows a steal phenomenon to the fistula (curved arrow) without filling the basilar artery. A T-shaped frame with detachable coils was positioned in both fistula and VA (D).
artery and used for control and diagnostic angiography. Two microcatheters (Excelsior\textsuperscript{1018}, Boston Scientific, Fremont CA) were navigated to fistulous portion at C1-2 junction. The first catheter tip was positioned in the vertebral artery just distal to fistulous hole. The second one is navigated into the fistulous hole. After making T-shaped anchoring frame with three detachable platinum coils successfully, subsequent packing with fibered coils was performed (Fig. 1D). Complete occlusion of the fistulous segment of the right vertebral artery and fistula itself was accomplished after total 19 coils were inserted. The post-embolization angiography showed complete occlusion of the right vertebral AVF and restoration of antegrade left vertebral artery (Fig. 1E & 1F).

The procedure was ended uneventfully. One day later, the patient’s symptom was relieved considerably. Follow-up MR images one month later demonstrated total disappearance of engorged epidural vein and normalized cervical cord without any abnormal signal intensity (Fig. 1H). The patient’s symptom was also relieved.

![Fig. 1. Final right (E) and left (F) vertebral angiographies depict complete occlusion of AVF and restoration of antegrade left VA flow. Follow-up MR image (H) one month later demonstrates complete disappearance of engorged epidural veins compressing spinal cord.](image-url)
DISCUSSION

Extracranial vertebral arteriovenous fistula (AVF) has a diversity of causes. Although traumatic cause, either accident or iatrogenesis, is more common, it can develop either spontaneous in origin. Various connective tissue diseases including neurofibromatosis, fibromuscular dysplasia, Marfan’s syndrome and Ehlers-Danlos syndrome are known to be predisposing factors (3, 4). In the evolution of the AVF in neurofibromatosis type 1 (NF-1), there are two possible mechanisms (3). Dysplastic smooth muscle or neurofibromatous proliferation itself in arterial wall could lead to aneurysmal formation, which leaks and ultimately ruptures into adjacent veins. Alternatively an arteriovenous fistula could develop congenitally as a manifestation of mesodermal dysplasia. The combination of cervical bony abnormality such as atlantoaxial dislocation may be another factor to develop this high flow fistula (5, 6). In present case, we could not see combined bony abnormality of the cervical spine.

More than 32 cases of spontaneous spinal AVF associated with NF-1 have been reported in the literatures (7). Reported patients ages ranged from 5 to 66 years (mean; 37.4). Most patients are female (84%). Radiculomyelopathy and bruit are common initial symptoms. The cause of the myelopathy are presumably due to compressed spinal cord by the dilated epidural venous channel or cord ischemia itself secondary to steal phenomenon or elevated venous pressure (8).

This disease entity must be separated from common dural spinal AVF type 1 which consists of a fistulous connection between a radicular artery and vein at the dural root sleeve with intradural drainage to the spinal cord venous system. In addition, unlike the dural AVF type 1, 97% of spontaneous spinal AVF associated with NF-1 originated from the vertebral artery in the cervical spine, none were found in the thoracic or sacral spine (4, 7).

Because vertebral AVF might have multiple feeding arteries, in the evaluation of vertebral AVF, full angiographies of possible arteries including bilateral internal and external carotid arteries, and all branches of both subclavian trunks are mandatory. The treatment of choice for vertebral AVF is complete obliteration of fistula. With the advent of endovascular techniques, occlusion with detachable balloons or coils can be a safe and feasible treatment method, which can avoid possible morbidity and mortality related to surgical treatment (7, 9-12). This patient underwent occlusion of fistula with detachable platinum coils and fibered coils. To secure embolization route of distal vertebral artery to the fistulous hole, we put two microcatheters. One is positioned in the fistulous hole and the other is in the vertebral artery just distal to fistula. After making T-shaped, stable frame positioned in both vertebral artery and fistula with detachable coils, we could insert push-type fibered coils. We were concerned about hyperperfusion syndrome due to abrupt restoration of vertebral blood flow. Long standing vascular steal from the intracranial circulation, resulting in tissue ischemia and loss of normal vascular autoregulation. On abrupt closure of the fistula, normal perfusion pressure breakthrough can result in neurologic dysfunction and/or hemorrhage (14, 15). After fistula obliteration combined with parent arterial occlusion, Strict blood pressure control was maintained during admission at the intensive care unit.

The presented case points out the complexity of clinical disorders encountered with NF-1, as it may show this kind of vascular manifestation in addition to well known and more common developed cutaneous, osseous and central nervous system.

In conclusion, we experience spontaneous vertebral AVF associated with NF-1 and treated with detachable coils and fibered coils successfully. The stability of the occlusion site is well confirmed by one month follow-up MR imaging.

References


Key Words: Neurofibromatosis; Vertebral arteriovenous fistula; Embolization