Rupture of a Cervical Arteriovenous Fistula Associated with Neurofibromatosis Type 1. A Case Report.

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ABSTRACT. We present a case report of a ruptured arteriovenous fistula (AVF) associated with neurofibromatosis type 1 (NF1). A 39-year-old man with NF1 suddenly developed a huge hematoma in the right neck region. Right vertebral and right extracarotid angiograms disclosed dilated, tortuous vessels and blood pooling. Therefore, superselective embolization using polyvinyl alcohol (PVA) particles and microcoils was performed to obliterate the AVF.

Key words: neurofibromatosis type 1 — arteriovenous fistula — transcatheter embolization — polyvinyl alcohol

Extracranial vertebral artery (VA) aneurysms and vertebral arterioveneous fistulas (VAVFs) are relatively rare disorders. The most frequent cause of these conditions is trauma, and atraumatic lesions are less common.[1-3] We describe a patient with NF1 in whom an AVF in the neck was treated with PVA particles and microcoils, with a favorable outcome.

CASE REPORT

A 39-year-old man experienced sudden right neck pain without any inducement. He also felt a tense feeling associated with a large subcutaneous mass in the right neck region. He was admitted to the hospital because of progressive dyspnea, and an emergency trachiostomy was performed.

In the childhood, the patient had been diagnosed as having NF1. On physical examination, there were multiple bean sized neurofibromas and a few cafe au lait spots on the body stem. He had no history of trauma to the neck or head or otolaryngologic disease, and no family history of vascular or neurological disorders. His blood pressure was 108/70 mmHg and laboratory tests did not show him to be anemic. He had no neurological abnormalities.

Computed tomography (CT) revealed a huge hematoma localized in the right neck region, but no evidence of a tumorous lesion (Fig 1). Therefore, emergency angiography was performed.

Right vertebral angiograms (Fig 2a, b) showed dilated, tortuous vessels, blood pooling, and dilated muscular branches arising from the C1/2 level. Superselective angiograms of each branch using a microcatheter clearly demonstrated the dilated, tortuous vessels and blood pooling that were seen in

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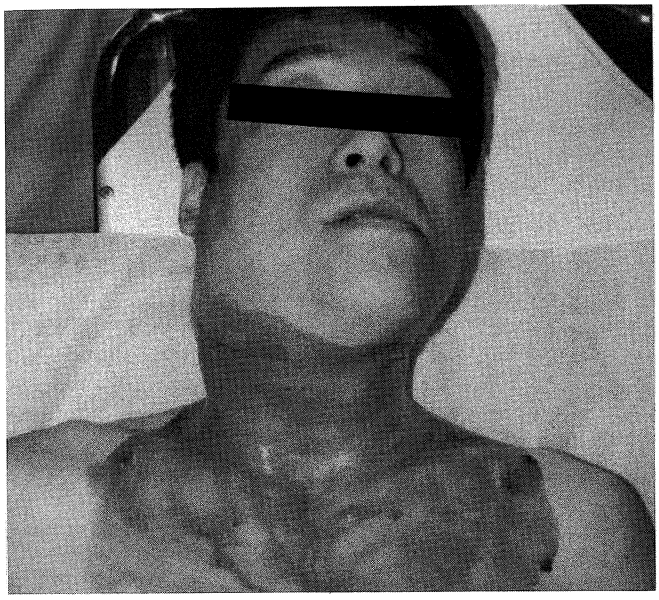
the vertebral angiograms (Fig 3a, b). The fistulous blood was drained into the sigmoid sinus. A diagnosis of extracranial arteriovenous fistulas with rupture was made.



Severe right neck swelling was seen.

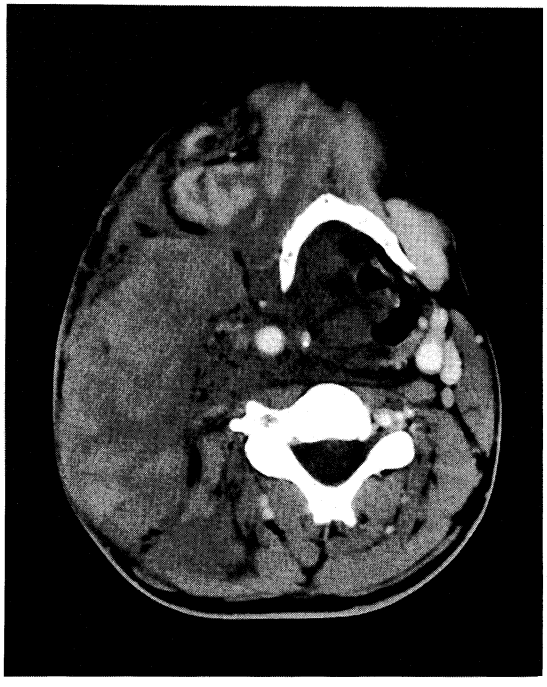


Fig 1. Computed tomography (CT) shows a huge hematoma in the right neck region.

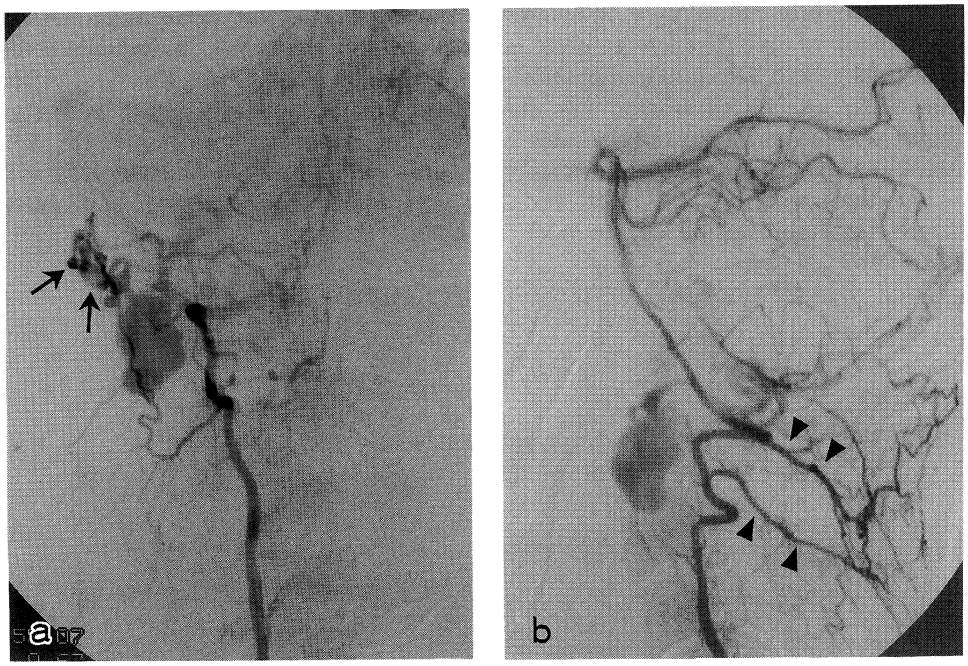


Fig 2a. A right vertebral angiogram, anteroposterior view, reveals dilated, tortuous vessels (arrow) and blood pooling.
Fig 2b. In a lateral view, two dilated muscular branches arising from the C1/2 level are noted (arrow heads).

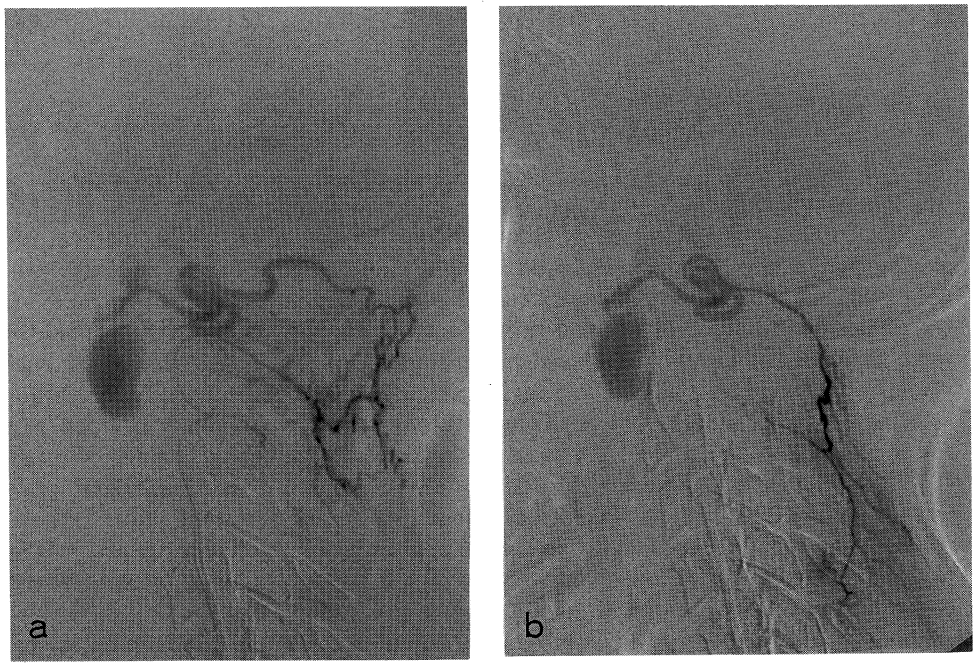


Fig 3a : Upper branch 3b : Lower branch
A superselective angiogram of each branch shows the tortuous vessels and blood pooling.

To reduce shunt flow, transarterial embolization was performed. After placement of a 6 French guiding catheter in the right VA, a microcatheter (Fastracker-18 : Target Therapeutics) was advanced coaxially within it. After the tip of the microcatheter was placed as distal as possible in the muscular branches, the anastomotic channels between the muscular branches of the VA were occluded with PVA particles (100 μm) (Mfg. by : Cook Incorporated). After this procedure, a right vertebral angiogram demonstrated apparently successful embolization of the AVF (Fig 4).

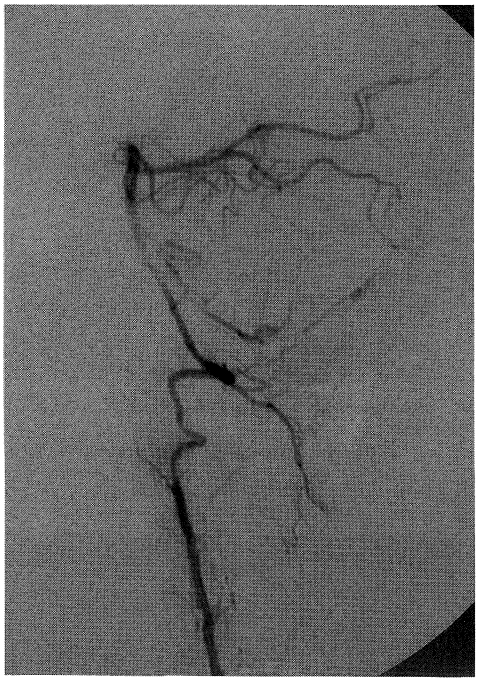


Fig 4. Post embolization, an angiogram shows complete obliteration of the fistula.

To confirm the presence of another flow to the AVF, right extracarotid angiography was performed, and vague abnormal blood pooling was demonstrated. This finding suggested the presence of crossflow to the AVF via the posterior auricular artery and an abnormal paracytic artery arising from the right extracarotid artery. Superselective angiography was performed for each vessel. A posterior auricular angiogram directly demonstrated the blood pooling that was seen before, and the fistulous blood was directly drained into the sigmoid sinus (Fig 5a). A microcatheter was advanced coaxially through the right extracarotid artery, and additional embolization was performed using microcoils (2 mm / 10 mm Complex Helical Fibered Platinum Coil : Target Theraupeutics). The paracytic artery arising from the extracarotid artery also contributed a tiny blood flow to the AVF (Fig 5b), and superselective embolization was performed using PVA particles (100 μm). After the procedure, right extracarotid angiography revealed complete obliteration of the fistula. No neurological abnormalities were observed during or after the operation.

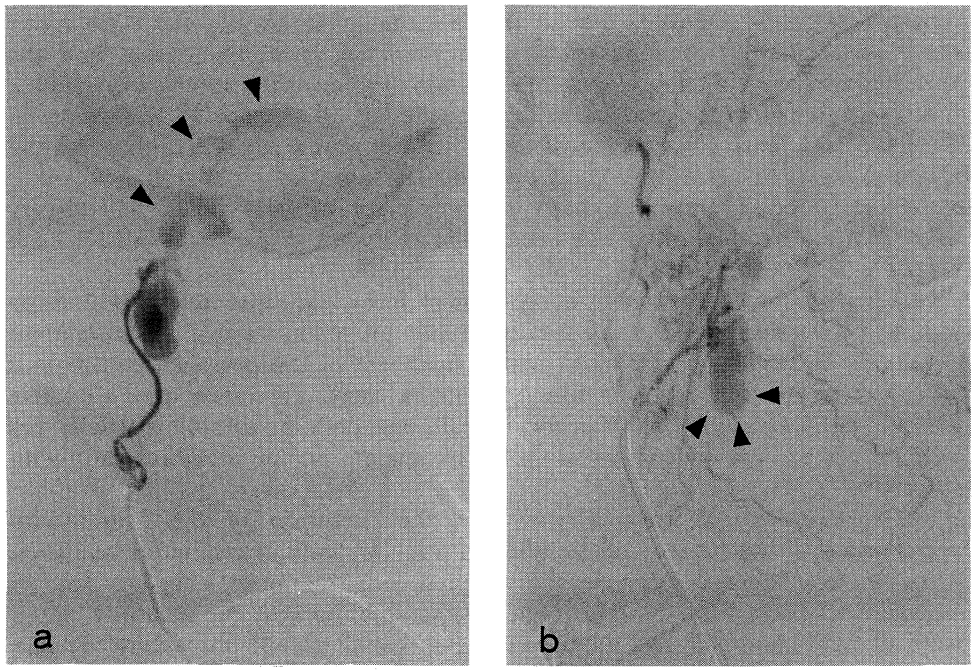


Fig 5a. A superselective posterior auricular angiogram directly demonstrates the blood pooling. The fistulous blood drained into the sigmoid sinus (arrow heads).
Fig 5b. A superselective angiogram of the paracytic artery arising from the right extracarotid artery shows a vague blood pooling (arrow heads).

After three weeks, the hematoma in the right neck was surgically removed, and the patient made an uneventful recovery with no subjective complaints.

DISCUSSION

NF1 (also known as von Recklinghausen's neurofibromatosis) is a hereditary disease characterized by tumors of the nervous system and dermal lesions as the main clinical symptons. It is sometimes complicated with osseous and endocrine abnormalities, but rarely with vascular lesions. Brasfield *et al*[4] found the frequency of vascular lesions to be 3.6 %. Vascular abnormalities could possibly arise in all the vessels, and many reports in which they have been given as a severe sympton from a rupture are accumulating.[5–8]

Generally, an AV fistula may occur secondary to neck trauma, such as may be encountered during chiropractic manipulation[9] or atlantoaxial dislocation,[10] or it may arise spontaneously without a clear history of trauma.[1–3]

The pathogenesis of fistula formation in NF1 is controversial. There are two possible mechanisms.[11] Either dysplastic smooth muscle or neurofibromatosis proliferation in the arterial wall could lead to aneurysm formation, leakage, and ultimately rupture into adjacent veins. Alternatively, an arterioveneous fistula could arise congenitally as a manifestation of mesodermal dysplasia.

Vascular abnormalities associated with NF1 are relatively frequent in vertebral arteries. More than 20 cases of vertebral AVF have been

reported.[1-3,8,10,12,13] This is due to the fact that anatomically there are many nerve roots arising from the spinal cord near the vertebral arteries, and the vertebral arteries have many anastomoses with the occipital artery, branches of the thyrocervical trunk, and muscular branches of all arteries on both sides of the neck. Our case could be consistent with the second mechanism because the CT scan did not reveal any clear tumorous lesions.

Surgical obliteration has been performed for cervical AVF, but surgery often has had to be aborted because the blood vessels of patients with NF1 are fragile and the surgical field is very vascular.[10,11]

Advances in endovascular neurosurgical techniques and tools have made less invasive treatment of many surgically difficult or inaccesible vasculopathies possible. The endovascular approach to treatment of the complex and difficult vascular lesions seen in neurofibromatosis may represent an attractive alternative to surgery. Use of occlusive balloons, coils or embolic particles has been reported previously as a solo treatment or in combination with surgery.[1-3,10-13]

The best way to treat a VAVF is the occlusion of the fistula with preservation of the patency of the affected VA. Usually, transarterial occlusion using a detachable balloon is the simplest and the most effective way of treating the direct fistula.[2] However, in this case, because the fistulous vessels were clearly identified and a microcatheter could placed into those vessels, superselective embolization using PVA particles and microcoils was selected to avoid the unexpected adverse events of complete occlusion of a vertebral artery.

PVA particles are unabsorbable embolic agents that are made in many sizes (50 ~ 2000 μm), and have been used to treat arteriovenous malformations for the past two decades.[14] However, to our knowledge, there has been only one reported case of treatment of VAVFs associated with NF1 using PVA particles. In the treatment of VAVFs using PVA particles, to prevent migration of the particles into the intracranial vertebral circulation because of overinjection pressure, the tip of catheter must be placed as distal as possible in the muscular branch, and the PVA particles must be suspended with radiographic contrast media, so that the operator can inject cautiously under roentgenological examination. Furthermore, anastomotic channels should be obliterated using relatively large particles to prevent flow of the particles into the venous circulation through the AV shunt. If there is a possibility of the particles flowing into drainage vessels, such as the posterior auricular vessel in this case, appropriate selection of embolic materials is very important.

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