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Case Report

Ruptured Occipital Artery Aneurysm Following Craniotomy in a Patient with Neurofibromatosis Type 1: Case Report

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Objective: Neurofibromatosis type 1 (NF1) is associated with vascular fragility, which results in aneurysms, arteriovenous fistulas, and dissections. Here, we describe a case of endovascular treatment of a ruptured occipital artery aneurysm that occurred after a craniotomy in a patient with NF1.

Case Presentation: A 46-year-old man with a history of NF1 underwent a right lateral suboccipital craniotomy to remove a cavernous hemangioma in the right middle cerebellar peduncle. Severe bleeding occurred in the occipital artery during the craniotomy. Due to vessel fragility, coagulation and ligation were not possible, and pressure hemostasis was achieved using cellulose oxide and fibrin glue. On postoperative day 12, the patient developed a sudden swelling on the right side of the neck as well as tracheal compression. Contrast-enhanced CT revealed a ruptured aneurysm in the right occipital artery. Transarterial embolization was performed under general anesthesia the same day. Right external carotid angiography showed an 18-mm-diameter fusiform aneurysm in the occipital artery. The aneurysm ruptured inferiorly to form a large pseudoaneurysm with significant jet flow. An arteriovenous fistula was also observed in a nearby vein. A microcatheter was inserted into the fusiform aneurysm under proximal blood flow control, and embolization was performed using coils and *N*-butyl-2-cyanoacrylate.

Conclusion: Compared to surgical repair of ruptured occipital artery aneurysms, endovascular treatment appears to be safe, effective, minimally invasive, and rapid. Ruptured occipital artery aneurysms in NF1 patients can cause neck swelling and airway compression and should be recognized as a potentially lethal condition.

Keywords neurofibromatosis type 1, occipital artery aneurysm, endovascular therapy, iatrogenic vascular injury

Introduction

Neurofibromatosis type 1 (NF1) is an autosomal dominant genetic disorder that occurs 1 in every 2000–3000 live births.[1)] Patients with NF1 have vascular fragility and vascular complications, including aneurysms, vascular dissection, and arteriovenous fistulas. Here, we describe a case of endovascular treatment for a ruptured occipital artery aneurysm that occurred after craniotomy in a patient with NF1 with a favorable outcome.

Case Presentation

A 46-year-old man with a history of NF1 was admitted to our hospital for surgery of a cavernous hemangioma. The cavernous hemangioma was located in the right middle cerebellar peduncle and associated with intratumoral bleeding (**Fig. 1**). A right lateral suboccipital craniotomy was performed. The skin and muscle incisions caused severe bleeding from a branch of the occipital artery. Attempts were made to stop the bleeding using coagulation and ligation; however, hemostasis was difficult to achieve because of vessel fragility and its tendency to tear rapidly. Additional inferior skin and muscle incisions were made to achieve hemostasis via compression with oxidized cellulose and fibrin glue. The cavernous hemangioma was subsequently removed as scheduled, and the patient's

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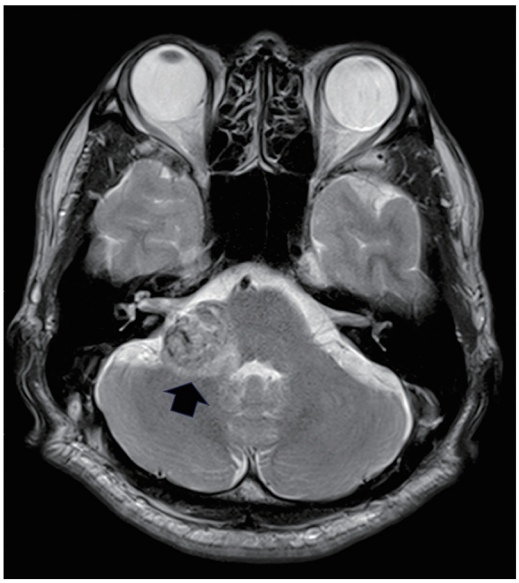

Fig. 1 Preoperative T2-weighted magnetic resonance image showing a cavernous hemangioma with surrounding edematous changes in the right middle cerebellar peduncle (black arrow) as well as an internal hemorrhage.

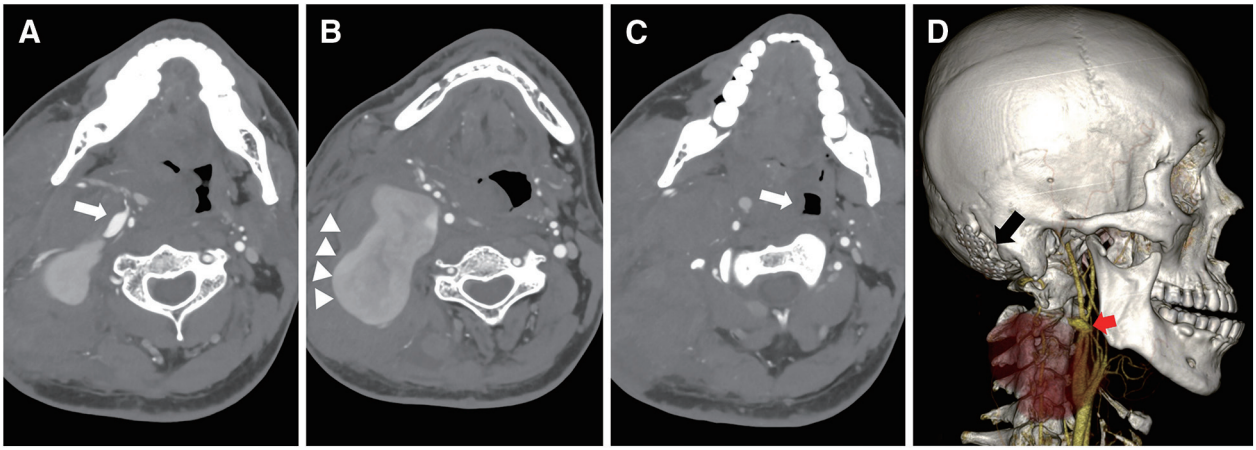

Fig. 2 Contrast-enhanced CT image showing a fusiform aneurysm in the occipital artery (**A**: white arrow). A large pseudoaneurysm protrudes inferiorly contiguous to the aneurysm (**B**: white arrowheads). The soft tissues of the neck are swollen due to bleeding, and the trachea is markedly deviated to the left (**C**: white arrow). Three-dimensional CT angiography reconstruction image (**D**) showing a fusiform aneurysm (red arrow) of the occipital artery at a lower position than the craniotomy (black arrow).

postoperative course was uneventful. At 12 days postoperative, the patient suddenly developed marked swelling on the right side of the neck and respiratory distress due to tracheal compression. Contrast-enhanced CT performed immediately thereafter revealed a fusiform aneurysm in the right occipital artery and abnormal vessel dilatation similar to a pseudoaneurysm. The soft tissues of the right neck were swollen, and the trachea was compressed and deviated to the left due to subcutaneous bleeding (**Fig. 2**).

Immediately after the tracheal intubation, the patient underwent endovascular treatment under general anesthesia. A 7F Optimo EPD guiding catheter (Tokai Medical Products, Inc., Aichi, Japan) with a balloon was placed in the right external carotid artery using the transfemoral approach. At the beginning of the procedure, 3000 units of unfractionated heparin were administered intravenously for systemic heparinization. The intraoperative whole-blood active clotting time was 200–250 seconds. Angiography of the right external carotid artery identified an 18-mm-diameter fusiform aneurysm in the right occipital artery. The aneurysm ruptured inferiorly to form a large pseudoaneurysm with significant jet flow. An arteriovenous fistula was also observed in a nearby vein. Blood flowed through the arteriovenous fistula into the internal

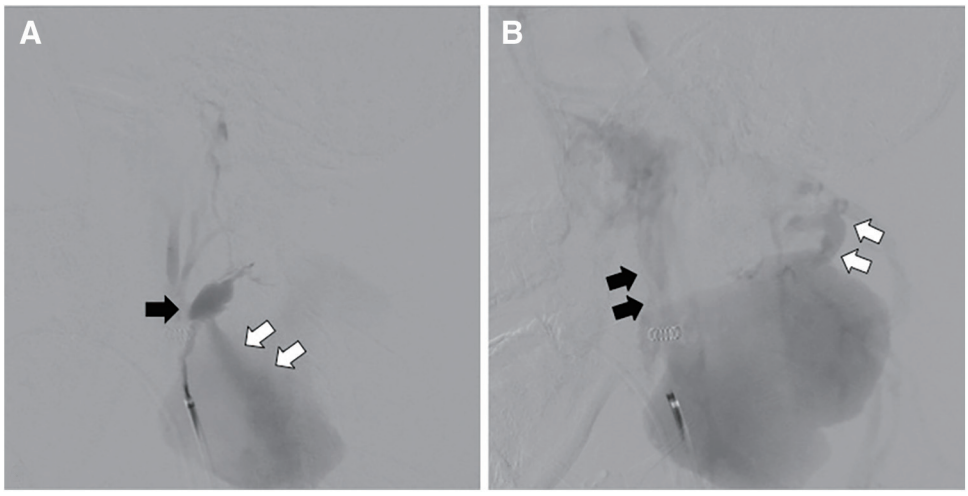

Fig. 3 (**A**) In the early arterial phase of right external carotid artery angiography, a fusiform aneurysm with an 18-mm diameter is visible in the proximal occipital artery (black arrow), which ruptured downward to form a large pseudoaneurysm (white arrows). (**B**) Right external carotid artery angiography in the late arterial phase showing the formation of an arteriovenous fistula with surrounding veins and blood draining into the vertebral venous plexus (white arrows) and internal jugular vein (black arrows).

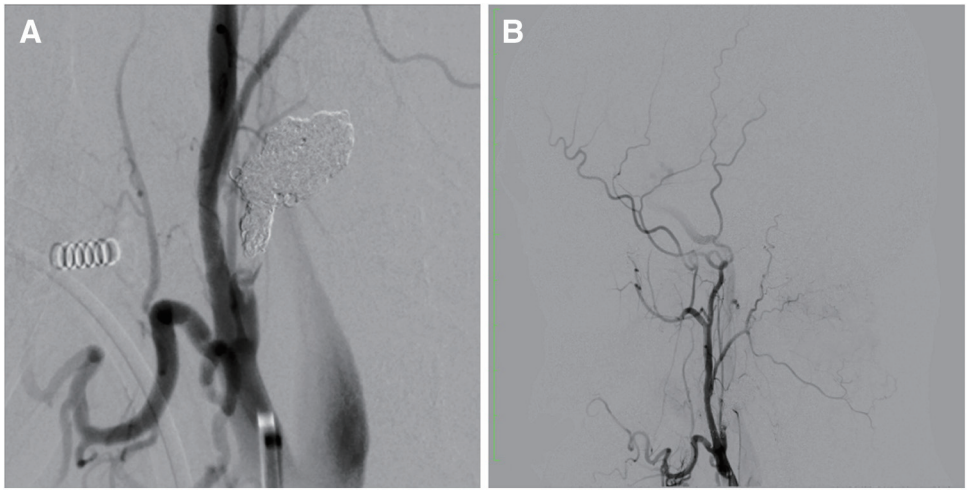

Fig. 4 (**A**) The fusiform aneurysm and proximal occipital artery were embolized with coils and *N*-butyl-2-cyanoacrylate. (**B**) Postoperative right external carotid artery angiography showing the disappearance of the aneurysm and arteriovenous fistula.

jugular vein and the vertebral venous plexus with intracranial reflux (**Fig. 3**).

When blood flow in the external carotid artery was interrupted by inflation of the balloon of the Optimo EPD catheter, an SL-10 microcatheter (Stryker, Kalamazoo, MI, USA) was inserted into the fusiform aneurysm. The fusiform aneurysm and proximal portion of the occipital artery were embolized using 17 detachable coils. After blocking the blood flow in the external carotid artery, external carotid angiography revealed a small amount of blood flow inferiorly from the aneurysm to the pseudoaneurysm. Due to the limited length of the occipital artery that could be embolized, embolization with 33% *N*-butyl-2-cyanoacrylate/lipiodol mixture was performed with the microcatheter positioned just proximal to the coil mass. Angiography performed after the embolization demonstrated complete occlusion of the fusiform aneurysm and disappearance of the pseudoaneurysm and arteriovenous fistula (**Fig. 4**). CTA performed on the day after the embolization showed the disappearance of the enhancement of the pseudoaneurysm and improved the neck swelling. The hematoma was not removed from the neck in the acute phase because the hematoma and pseudoaneurysm could not be identified through the body surface. The need for

Table 1 Summary of nine cases of ruptured occipital artery aneurysms associated with neurofibromatosis type 1

Author	Year	Age	Sex	Symptom	Tracheal intubation	Tracheotomy	Treatment	Embolic material	Outcome
Kanematsu et al.[10]	2011	48	M	Neck pain, neck swelling, dyspnea	No	–	TAE	Coil	Survived
Kanematsu et al.[10]	2011	39	F	Neck pain, neck swelling	No	–	TAE	Coil	Survived
Ando et al.[11]	2023	40	F	Neck pain, neck swelling, dyspnea	Failure	Yes	TAE, surgery	Coil	Survived
Imahori et al.[12]	2016	70	F	Neck pain, neck swelling	No	–	TAE	Coil, NBCA	Survived
Bissacco et al.[13]	2018	53	F	Neck swelling	Yes	–	TAE, surgery	Particle, NBCA	Survived
Takeda et al.[14]	2010	48	M	Neck swelling, dyspnea	Failure	Yes	TAE	Coil	Survived
Takeda et al.[15]	2002	52	M	Neck swelling, dyspnea	Yes	–	TAE	Coil	Dead
Takeda et al.[15]	2002	60	F	Neck swelling	No	–	TAE	Coil	Survived
Present case	2023	46	M	Neck swelling, dyspnea	Yes	–	TAE	Coil, NBCA	Survived

NBCA: *N*-butyl-2-cyanoacrylate; TAE: transarterial embolization

removal was considered after approximately 1 week of follow-up, but the neck swelling improved immediately. Thus, the hematoma was not removed. Moreover, a tracheotomy was not performed, and the patient was extubated 2 days after embolization.

Discussion

NF1 is a dominantly inherited genetic disorder resulting from germline mutations in the *NF1* tumor suppressor gene. In addition to skin lesions, such as café-au-lait spots, neurofibromas, plexiform neurofibromas, skull dysplasia, and brain tumors are typical lesions.[1]

NF1 has been associated with vascular fragility and complications of vascular diseases, such as aneurysms, dissection, vascular rupture, and arteriovenous fistulas. The frequency of this vascular complication is reportedly 0.4%–6.4%.[2–4] However, these vascular lesions may be underestimated because of their potential to remain undiagnosed. The mechanism of this vascular fragility in the arteries is considered the dense proliferation of spindle cells within the arterial walls accompanied by medial cystic degeneration, disruption of the tunica elastica, and ischemia of the vessels themselves due to compression of the nutrient vessels by tumor cell infiltration.[5–7] In the veins, the direct infiltration of neurofibromatous cells into the vessel wall also causes fragility.[8] However, these fragile changes do not occur in all vessels of patients, and it is difficult to distinguish between normal and affected vessels.[4] The spontaneous rupture of vessels that do not form aneurysms has also been reported.[9] The most commonly affected vessels are the intercostal and subclavian arteries. Although the vascular invasion of tumor cells is considered a possible mechanism, it can cause sudden hemorrhagic complications that cannot be predicted in advance and are sometimes fatal.

There have been many reports of aneurysms associated with NF1 in vessels throughout the body, including intracranial and extracranial vessels, and many have a severe course due to critical bleeding. Although iatrogenic occipital artery aneurysms associated with NF1 have not been previously reported, there have been eight cases of ruptured idiopathic occipital artery aneurysms associated with NF1.[10–15] A summary of these nine cases, including the present case, is presented in **Table 1**. Five of the nine patients had airway emergencies and were intubated. In two cases, intubation was impossible, and an emergency tracheostomy was performed. All patients underwent endovascular treatment, and two patients underwent direct surgery. One patient died. This is a fatal condition with a high probability of airway emergency due to bleeding. A high degree of airway compression may make normal tracheal intubation impossible, thus requiring a tracheostomy. In the present case, inadequate hemostasis of the injured occipital artery during craniotomy may have resulted in the formation of a fusiform aneurysm that ruptured during the subacute postoperative period.

The vascular fragility of patients with NF1 may result in bleeding from sites that are not directly visible in the operative field and cannot be managed intraoperatively. In such cases, an early postoperative vascular evaluation and repeated follow-up should be performed with delayed bleeding in mind. In this case, an arteriovenous fistula also formed in the surrounding veins. The mechanism of this arteriovenous fistula is thought to be the collapse of the surrounding fragile vein wall due to pressure damage caused by the rupture of the aneurysm, resulting in the formation of an arteriovenous fistula. Two previous reports of NF1-associated occipital artery aneurysm rupture showed an arteriovenous fistula of the internal jugular vein, similar to that in the present case.[12,15)]

In cases of traumatic or iatrogenic arterial rupture or arteriovenous fistulas, such as in the present case, the rupture or shunt point is often a solitary lesion that can be cured by targeted embolization. However, high-flow shunts may occur, which makes it challenging to accurately identify the rupture point and control the catheter. The use of a balloon-guiding catheter facilitates angiography and treatment under controlled blood flow conditions with proximal interruption.

Conclusion

Here, we reported a rare postoperative complication of occipital aneurysm rupture in a patient with NF1. Occipital artery aneurysm rupture is an airway emergency and potentially fatal complication that requires careful attention during craniotomy and postoperative management. Endovascular trapping with proximal flow control was effective for ruptured iatrogenic pseudoaneurysms of the occipital artery with a high-flow arteriovenous shunt.

Disclosure Statement

The authors declare that they have no conflicts of interest.

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