



Case Report

Traumatic arteriovenous fistula of the superficial temporal artery caused by massive subcutaneous hematoma prompting surgical removal and endovascular treatment in a patient with neurofibromatosis type 1

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ABSTRACT

Background: Neurofibromatosis type 1 (NF-1) is often characterized by vascular disorders related to vessel vulnerability that can lead to unfavorable outcomes. Here, we describe a case of NF-1 complicated with a massive subcutaneous hematoma posing a risk of visual impairment for which rapid decompression and a subsequent less invasive approach result in a favorable outcome.

Case Description: A 40-year-old woman with NF-1 presented with a massive left subcutaneous temporal hematoma following a mild head contusion. Four days after hospitalization, the hematoma increased in size and severely compressed the left eye, prompting immediate hematoma removal to preserve visual function. Immediately after the hematoma removal, a superficial temporal arteriovenous fistula was found on the digital subtraction angiography and embolized by the endovascular procedure. Her visual acuity was preserved, and no bleeding recurrence was observed throughout the follow-up.

Conclusion: Surgical hematoma removal followed by endovascular treatment was effective in preserving visual function. Since vessel fragility is characteristic of patients with NF-1, it should be kept in mind that vascular complications may lead to serious clinical outcomes. In certain NF-1 cases, less invasive treatments for vascular abnormalities may be preferable.

Keywords: Endovascular treatment, Neurofibromatosis type 1, Subcutaneous hematoma, Superficial temporal artery, Traumatic arteriovenous fistula

INTRODUCTION

Neurofibromatosis type 1 (NF-1) is an autosomal dominant disease occurring in 1 of 3000–4000 individuals with characteristic manifestations of café-au-lait spots, neurofibromas, and Lisch nodules.^[4] The major causes of death are malignant neoplasms and vascular disorders, including vessel dissections, aneurysms, arteriovenous (AV) malformations, and arteriovenous fistulas (AVFs), with the estimated

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vasculopathy rate of 0.4–6.4%.^[3,11] Several mechanisms of vessel vulnerability have been proposed,^[6,11] although they are still not fully understood. Here, we report a massive subcutaneous hematoma that resulted in the formation of a superficial temporal artery (STA) AVF in a patient with NF-1.

CASE REPORT

A 40-year-old woman with NF-1 with a history of optic pathway/hypothalamic glioma underwent surgery with the right eye removal at the age of 3. She recently hit her left temple slightly on the corner of a PC monitor. Three days later, that area suddenly swelled, and she visited our hospital. She was alert without a neurological deficit. Head computed tomography (CT) showed a marked subcutaneous hematoma on the left [Figure 1a]. The blood test, chest X-ray, and electrocardiogram were normal. Digital subtraction angiography (DSA) showed poorly contrasted distal left STA due to compression by hematoma in the early arterial phase of the left external carotid artery (ECA) on admission [Figure 1b and c] and minor extravasation near the contusion site in a late arterial phase [Figure 1d]. Since the diameter of the vessel involved in the extravasation was too small for microcatheter navigation, and the hematoma did not enlarge over time, we expected it to resolve spontaneously.

Four days after admission, the hematoma suddenly enlarged, preventing the patient from opening her left eye, and intraocular pressure increased to the point of causing blindness. Head CT revealed left eye deformation due to compression by the enlarging hematoma [Figure 2a]. Thus, we performed surgical hematoma removal under general anesthesia to avoid bilateral blindness. After a small skin incision near the left temple, the hematoma was partially removed. However, no obvious bleeding point was detected, and the bleeding oozing from undefined locations was difficult to control. Therefore, we performed an endovascular intervention, considering it more favorable than surgery in terms of accuracy of vascular pathology assessment and avoiding excessive bleeding. A long 4 Fr sheath was placed in the right femoral artery, and a 4-Fr

JB-2 catheter was advanced to the left ECA. The angiography showed a string sign in the left proximal STA, suggesting arterial dissection [Figure 2b]. It was connected to a remarkably dilated vein (varix) draining to the facial and external jugular veins through the fistula [Figure 2b and c]. We replaced the 4-Fr sheath with a 6-Fr one, and a 6-Fr guiding catheter was advanced into the left ECA. Excelsior 1018 catheter (Stryker, Fremont, CA, USA) was steered just proximal to the STA dissection point, where a Target US 360 3 mm × 8 cm coil (Stryker, Fremont, CA, USA) was then placed with several additional soft coils to embolize the proximal portion of the STA. As a result, the varix, AV shunt, and drainage veins were blocked [Figure 3a and b]. Follow-up CT showed recovery of the left eye deformity without rebleeding, and the vision was preserved. Two months later, follow-up angiography confirmed no vascular abnormality recurrence [Figure 3c and d].

DISCUSSION

We treated a patient with NF-1 effectively using a combination of surgical decompression and endovascular treatment. In NF-1, hemorrhagic complications may be associated with vessel vulnerability. Mild injuries can lead to unfavorable outcomes, such as a life-threatening brachial artery hemorrhage requiring amputation of the upper extremity^[8] or a massive hemothorax without any traumatic episodes.^[10] Three hypotheses of vessel fragility mechanisms in NF-1 have been discussed: neurofibromas around vessels invade arterial walls, resulting in their weakening; an increased number of spindle cells in the vascular smooth muscle layer leads to vessel wall fragility; and neurofibromas exert compression on vascular nutrient vessels.^[6,11] To date, there have been several reports of perioperative severe hemorrhagic complications.^[1,8,10] Raborn *et al.* demonstrated the efficacy of endovascular treatment of hypertension caused by renal artery stenosis in two pediatric patients with NF-1.^[11] Massive subcutaneous bleeding from a non-traumatic STA pseudoaneurysm in a patient with NF-1 was successfully treated using an endovascular approach.^[7] In our case, a mild head contusion resulted in a massive temporal

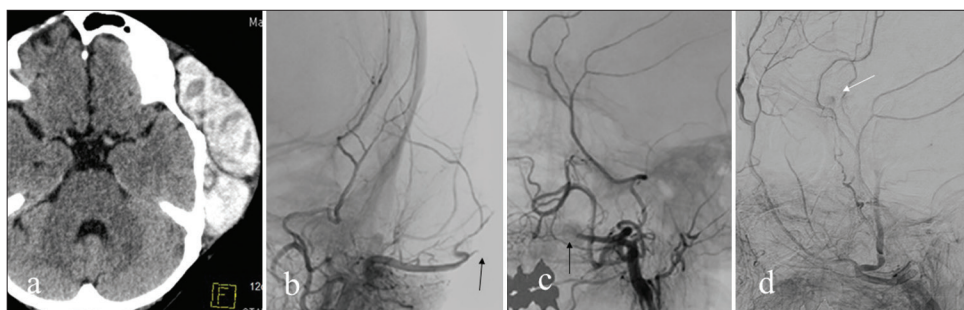


Figure 1: (a) Head CT: a massive left subcutaneous hematoma on admission. (b) AP and (c) lateral views of the early arterial phase external carotid artery (ECA) digital subtraction angiography (DSA): poorly contrasted distal left STA due to compression by hematoma (black arrow). (d) The lateral view of the late arterial phase ECA DSA: minor extravasation near the contusion site (white arrow).

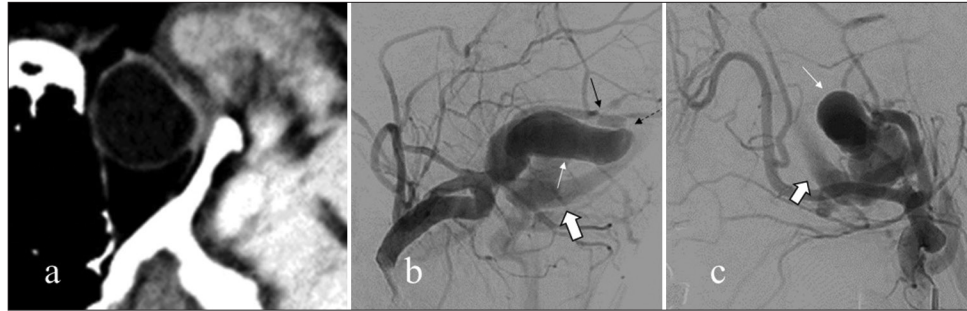


Figure 2: (a) Axial computed tomography. (b) Anterior- posterior view of the left external carotid artery (ECA) digital subtraction angiography (DSA): string sign of the left STA (black arrow), the dilated vein (white arrows), and the vein (bold arrows) draining via the arteriovenous fistula (dotted arrow). (c) Lateral view of the left ECA DSA: the dilated vein (white arrows) and the vein (bold arrows) draining via the arteriovenous fistula.



Figure 3: (a) Anterior- posterior and (b) lateral views of the left external carotid artery (ECA) digital subtraction angiography (DSA) immediately after embolization. (c and d) Follow-up of DSA of the left ECA two months after the intervention.

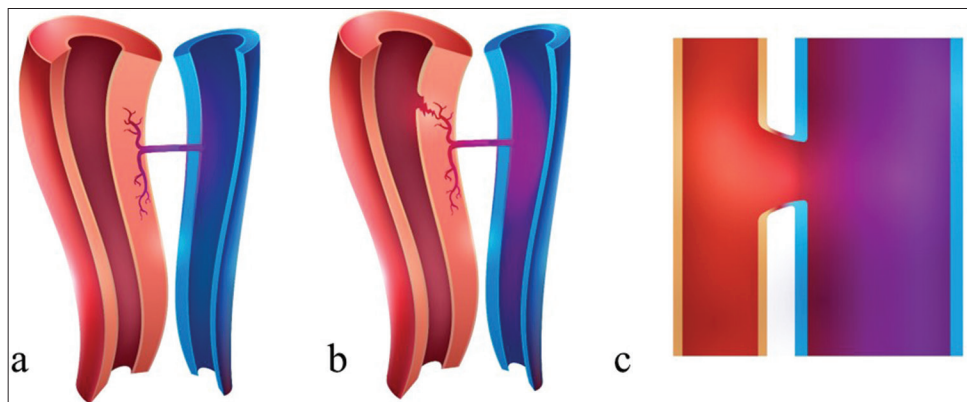


Figure 4: (a) Illustrations show *venous vasa vasorum* and a hypothetical mechanism of AV shunt formation. (b and c) The STA stretching by the massive subcutaneous hematoma caused arterial wall laceration that could reach *venous vasa vasorum*, resulting in STA-STV shunt formation.

subcutaneous hematoma, and DSA demonstrated AVF between the STA dissection and the varix. These vascular abnormalities were eliminated, and severe hemorrhagic complications were avoided with our approach.

The laceration theory has been proposed as a mechanism of traumatic AVF formation.^[2,9] Simultaneous laceration of the artery and the accompanying vein may have resulted in a single direct connection between the vessels. However, the

uniqueness of the present case was that the AV shunt point (the proximal STA near the zygomatic arch) was far from the head contusion site and the massive bleeding point (the distal STA near the left temple). It remains questionable that simultaneous laceration of the proximal STA and accompanying vein could have occurred without direct traumatic impact or bleeding. Anatomically, there are *vasa vasorum*, which feeds blood vessel walls, and *venous vasa vasorum*, which originate from the

arterial wall and drain into the main lumen or branches of the accompanying veins [Figure 4a].^[5,12] Here, we hypothesize that the STA stretching by the massive subcutaneous hematoma caused arterial wall laceration that could reach *venous vasa vasorum* [Figure 4b], resulting in STA-superficial temporal vein shunt formation [Figure 4c].

In NF-1, attention should be paid to vessel vulnerability, which can lead to serious consequences. Our case may be instructive in terms of hemorrhagic complications, where emergent and less invasive interventions proved favorable.

CONCLUSION

We report a case of a massive subcutaneous hemorrhage that caused STA AVF formation in a patient with NF-1. Surgical hematoma removal followed by endovascular treatment was effective in preserving visual function. Since vessel fragility is characteristic of patients with NF-1, it should be kept in mind that vascular complications may lead to serious clinical outcomes. In certain NF-1 cases, less invasive treatment of vascular abnormalities might be favorable.

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Ethical approval

The Institutional Review Board approval is not required.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Nil.

Conflicts of interest

There are no conflicts of interest.

Use of artificial intelligence (AI)-assisted technology for manuscript preparation

The authors confirm that there was no use of artificial intelligence (AI)-assisted technology for assisting in the

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REFERENCES

- Bhaskar S, Sobti S, Singh AK. Massive scalp hematoma due to diffuse neurofibroma in NF-1. *Clin Neurol Neurosurg* 2013;115:477-8.
- Biegaj E, Rutkowska-Zimirka J, Radzymińska-Maliszewska M, Zaremba A, Pniewski J. Arteriovenous fistula of superficial temporal vessels. *Folia Morphol (Warsz)* 2019;78:879-82.
- Chew DK, Muto PM, Gordon JK, Straceski AJ, Donaldson MC. Spontaneous aortic dissection and rupture in a patient with neurofibromatosis. *J Vasc Surg* 2001;34:364-6.
- Evans DG, Howard E, Giblin C, Clancy T, Spencer H, Huson SM, et al. Birth incidence and prevalence of tumor-prone syndromes: estimates from a UK family genetic register service. *Am J Med Genet A* 2010;152A:327-32.
- Gössl M, Rosol M, Malyar NM, Fitzpatrick LA, Beighley PE, Zamir M, et al. Functional anatomy and hemodynamic characteristics of vasa vasorum in the walls of porcine coronary arteries. *Anat Rec Part A* 2003;272A:526-37.
- Greene JF, Fitzwater JE, Burgess J. Arterial lesions associated with neurofibromatosis. *Am J Clin Pathol* 1974;62:481-7.
- Iwama T, Mizutani K, Kubo H, Katsumata M, Akiyama T, Toda M. A case of neurofibromatosis type 1 diagnosed after idiopathic rupture of superficial temporal artery pseudoaneurysm requiring endovascular treatment. *NMC Case Rep J* 2023;10:125-30.
- Lee J, Kim Y. Life-threatening brachial artery hemorrhage and a lethal outcome in patients with neurofibromatosis type 1: Two case reports and a review of the literature. *J Int Med Res* 2021;49:03000605211025344.
- Li F, Zhu S, Liu Y, Chen Y, Chi L, Chen G, et al. Traumatic arteriovenous fistula of the superficial temporal artery. *J Clin Neurosci* 2007;14:595-600.
- Negreira KE, Lichtenberger JP, Allais B, Alhaddad A, Bernetch M, Jain V. Subclavian artery branch pseudoaneurysm rupture with massive hemothorax in a patient with neurofibromatosis type 1. *Chest* 2020;157:e103-5.
- Raborn J, McCafferty BJ, Gunn AJ, Moawad S, Mahmoud K, Abdel AA, et al. Endovascular management of neurofibromatosis type I-associated vasculopathy: A case series and brief review of the literature. *Vasc Endovascular Surg* 2020;54:182-90.
- Shoenenberger F, Mueller A. On the vascularization of the bovine aortic wall. *Halvet Physiol Pharmacol Acta* 1960;18:13650.

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