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Case Report

Therapeutic challenges in transcatheter arterial embolization for an enlarging subcutaneous hematoma in a patient with neurofibromatosis type 1: A case report☆,☆☆

Keisuke Suzuki, M.D., Ph.D.[a,b,*], *Ryozo Kai, M.D.*[a], *Jiro Munechika, M.D., Ph.D.*[a], *Koji Morita, M.D.*[a], *Maya Makita, M.D.*[a], *Miho Saeki, M.D.*[a], *Takahiro Kanai, M.D.*[a], *Kenji Dohi, M.D., Ph.D.*[b], *Noritaka Seino, M.D., Ph.D.*[a,c], *Yoshimitsu Ohgiya, M.D., Ph.D.*[a]

[a] *Department of Radiology, Division of Radiology, Showa University School of Medicine, 1-5-8 Hatanodai, Shinagawa-ku, Tokyo 142-8666, Japan*

[b] *Department of Emergency and Disaster Medicine, Showa University School of Medicine, 1-5-8 Hatanodai, Shinagawa-ku, Tokyo 142-8666, Japan*

[c] *Department of Radiology, Showa University Koto Toyosu Hospital, 5-1-38 Toyosu, Koto-ku, Tokyo 135-8577, Japan*

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ABSTRACT

A 60-year-old woman with a history of neurofibromatosis type 1, who was admitted with pulmonary hypertension, developed buttock pain and anemia, and contrast-enhanced computed tomography showed a large subcutaneous hematoma with minimal active extravasation. Angiography of the bilateral internal iliac arteries revealed diffuse, irregular blood vessels without extravasation. As the exact bleeding site could not be identified, the patient was managed conservatively. However, the patient's symptoms and anemia worsened the following day. Repeat angiography revealed two pseudoaneurysms in the right inferior gluteal artery, which were embolized using n-butyl-2-cyanoacrylate. Nonetheless, the patient's anemia further worsened the following day. Repeat contrast-enhanced CT revealed another site of extravasation in the enlarging hematoma, but no extravasation was observed on the subsequent angiography. Owing to the worsening anemia and enlarging hematoma, proximal embolization of the irregular bilateral inferior gluteal arteries was performed using gelatin sponge particles. The patient's anemia and symptoms improved. Vasculopathy associated with neurofibromatosis type 1 is rare, with an incidence of approximately 3%. In patients with neurofibromatosis type 1, the blood vessels become fragile because of tunica media thinning and elastic-lamina rupture. Histopathologically, neurofibromatosis type 1-associated vasculopathy is characterized by a mixture of normal and abnormal vessels.

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* Correspondence: Keisuke Suzuki, M.D., Ph.D., Department of Emergency and Disaster Medicine, Showa University, 1-5-8 Hatanodai, Shinagawa-ku, Tokyo 142-8666, Japan. Tel: 81-3-3784-8000, Fax: 81-3-3784-8358
E-mail address: k.s.07202251@gmail.com (K. Suzuki).
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Abnormally fragile blood vessels may repeatedly rupture followed by physiological hemostasis, which may explain the diagnostic and therapeutic challenges during angiography in this case. In patients with neurofibromatosis type 1 with acute bleeding, irregular vessels without active extravasation on angiography may be indicated for embolization.

Introduction

Neurofibromatosis type 1 (NF-1) is an autosomal dominant genetic disorder with an incidence of approximately 1:3000–4000 individuals [1,2]. Also known as von Recklinghausen's disease, NF-1 is caused by a genetic mutation in the long arm of chromosome 17 [3]. NF-1 is characterized by skin lesions, such as café-au-lait spots and neurofibromas, and hemorrhage due to vascular fragility has been reported in few cases [4,5]. Bleeding is often difficult to stop and can be fatal. Although the potential complications of asymptomatic NF-1 - associated vascular lesions are unclear, the mean age at death in patients with NF-1 is 15.7 years lower than that in the general population [6].

However, consensus on angiographical, surgical, or conservative treatment strategies is lacking. Herein, we report a case of a large subcutaneous hematoma caused by minor trauma that was difficult to treat with endovascular therapy.

Case Report

A 60-year-old woman with a history of NF-1 was admitted with pulmonary hypertension. The patient was not taking any anticoagulants or antiplatelet medications, and blood tests revealed no abnormalities in her coagulation system (Table 1). She suddenly developed buttock pain and anemia after sitting in bed in the hospital room. Contrast-enhanced computed tomography (CT) revealed a large subcutaneous hematoma with small active extravasation (Fig. 1). The treatment of choice was transcatheter arterial embolization (TAE) using the microcoaxial catheter technique under local anesthesia and intravenous sedation. Angiography of the bilateral internal iliac arteries revealed diffuse irregular blood vessels without extravasation (Fig. 2). As the exact bleeding site could not be identified, the patient was managed conservatively. However, the patient's symptoms and anemia worsened on Day 2. Repeat angiography revealed two pseudoaneurysms in the right inferior gluteal artery, which were embolized using n-butyl-2-cyanoacrylate (Fig. 3). Nonetheless, the patient's anemia further worsened on Day 3. Repeat contrast-enhanced CT revealed another site of extravasation in the enlarging hematoma, but no extravasation was observed on the subsequent angiography. Owing to the worsening anemia and enlarging hematoma, embolization of the irregular bilateral inferior gluteal arteries was performed using gelatin sponge particles (Fig. 4), which improved the patient's anemia and symptoms.

Table 1 – Laboratory data on arrival

Laboratory Data	
WBC (/μL)	5,500
Hb (g/dL)	12.0
Plt (/μL)	308,000
BUN (mg/dL)	19.4
Cre (mg/dL)	0.42
AST/ALT (U/L)	21/14
CRP (mg/dL)	0.10
PT-INR/APTT	1.07/27.9
D-dimer (μg/dL)	1.11

WBC, white blood cell; Hb, hemoglobin; Plt, platelet; BUN, blood urea nitrogen; Cre, creatinine; AST, aspartate aminotransferase; ALT, alanine aminotransferase; CRP, C-reactive protein; PT-INR, prothrombin time-international normalized ratio; APTT, activated partial thromboplastin time.

Discussion

We encountered a therapeutic challenge in TAE for an enlarging subcutaneous hematoma in a patient with NF-1. Three consecutive days of examination and treatment were required for successful management. This treatment difficulty was attributed to the vascular fragility characteristic of NF-1.

The incidence of vascular involvement in NF-1 is 3.6% [7]. NF-1-associated vasculopathy affects large and small arteries, veins, and pulmonary arteries and can result in vascular stenosis, occlusion, aneurysms, pseudoaneurysms, rupture, or fistula formation [8]. Our patient was initially hospitalized with pulmonary hypertension, which is a rare but serious complication of NF-1 [9].

Possible mechanisms of NF-1-related hemorrhage include weakening of the vessel wall due to direct infiltration of the neurofibrillary tangles, weakening of the wall associated with ischemia due to compression of the nutrient vessels of the neurofibrillary tangles, and weakening of the elastic plate due to spindle-cell proliferation in the vascular intima [10,11].

Hemorrhage due to a ruptured artery can easily result in hemorrhagic shock, and a rapid response is important. The main treatment options for vascular lesions are surgery and TAE. TAE may be the first choice of treatment, because surgery is potentially dangerous and has a high mortality rate [5,12].

Misao et al. reported repeated hemostasis for spontaneous hemothorax, in which a new aneurysm appeared during follow-up [5]. This may have been due to vessel fragility. The findings in this case suggest that in patients with NF-1, even if extravasation appears to have disappeared, it is only

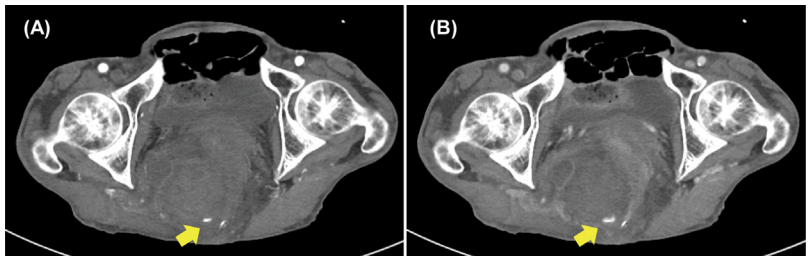

Fig. 1 – Pre-treatment axial contrast-enhanced CT images show a large subcutaneous hematoma with a small amount of active extravasation (arrow). (A) 30 s after injection of contrast medium. (B) 80 s after injection of contrast medium. CT, computed tomography

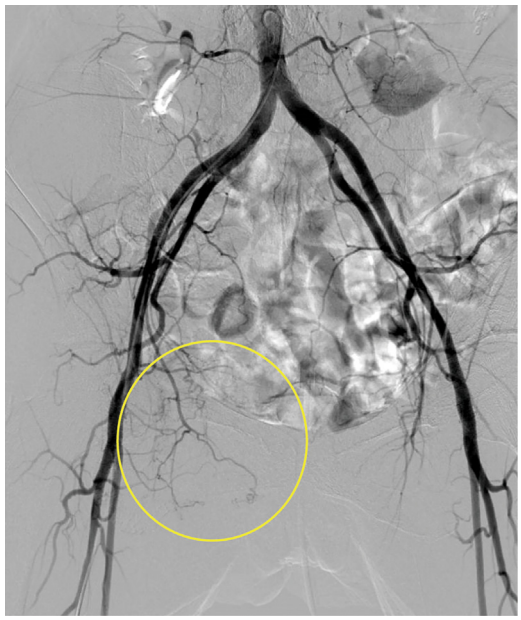

Fig. 2 – Angiography on Treatment Day 1
Angiography of the bilateral internal iliac arteries shows diffuse irregular blood vessels (circles) without extravasation. The exact bleeding site cannot be identified.

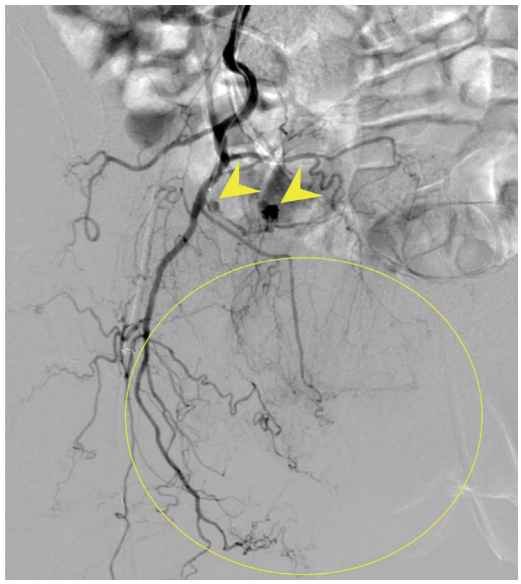

Fig. 3 – Angiography on Treatment Day 2
Repeat angiography demonstrates two pseudoaneurysms (arrowheads) in the right inferior gluteal artery and diffuse irregular blood vessels (circle).

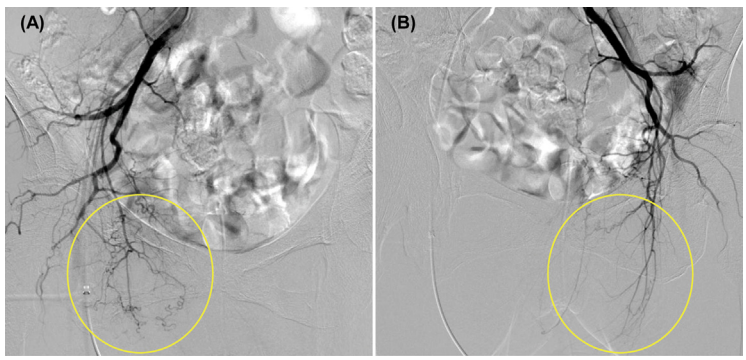

Fig. 4 – Angiography on Treatment Day 3 (A) Angiography of the right internal iliac artery. (B) Angiography of the left internal iliac artery.
No extravasation is observed.

invisible owing to vasospasm, and bleeding may recur. Therefore, in patients with NF-1 with acute hemorrhage, embolization may be indicated even if only irregular vessels without extravasation are observed on angiography.

Conclusion

NF-1 vasculopathy presents diagnostic and therapeutic challenges when active bleeding is not detected by angiography. In patients with NF-1 with acute bleeding, irregular vessels without active extravasation on angiography may be indicated for embolization.

Patient consent

Written consent was obtained from the patient, and patient's personal information was carefully anonymized with great care. The patient provided informed consent for publication of her case history.

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