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Case Report

Iliofemoral Aneurysm in Patients with Type 1 Neurofibromatosis: A Case Report and a Literature Review

Takeshi Uzuka, MD, PhD, Riko Umeta, MD, Hiroki Uchiyama, MD, PhD, Ayaka Arihara, MD, Hitoki Hashiguchi, MD, and Akihiko Sasaki, MD, PhD

Vascular involvement, especially in the iliofemoral segment, is rare in type 1 neurofibromatosis. We herein report a case involving a 49-year-old male diagnosed with type 1 neurofibromatosis who presented with right inguinal pain and swelling. CT angiography revealed a 50-mm aneurysm extending from the right external artery to the common femoral artery. Although surgical reconstruction was performed successfully, the patient required an additional operation 6 years later for aneurysm enlargement in the deep femoral artery. Histopathological examination confirmed the proliferation of neurofibromatosis cells in the aneurysm wall.

Keywords: type 1 neurofibromatosis, iliofemoral artery aneurysm

Introduction

Type 1 neurofibromatosis (NF1), also known as von Recklinghausen's disease, is a well-recognized genetic disorder characterized via abnormal cutaneous pigmentation (Café-au-lait spots) and multiple skin tumors. While its vascular involvement is known, the occurrence is infrequent.[1] Aneurysm formation in the iliofemoral artery is rare and, to our knowledge, has not been reported elsewhere. Therefore, we herein report a case involving an iliofemoral artery aneurysm that required a repeat operation to address aneurysm progression.

Cardiovascular Surgery, Sunagawa City Medical Center, Sunagawa, Hokkaido, Japan

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Corresponding author: Takeshi Uzuka, MD, PhD. Cardiovascular Surgery, Sunagawa City Medical Center, W4N3 1-1, Sunagawa, Hokkaido 073-0164, Japan
Tel: +81-125-54-2131, Fax: +81-125-54-0101
E-mail: Uzuka@aol.com

Case Report

A 49-year-old male diagnosed with NF1 was referred to our hospital for tenderness in the right groin. Enhanced CT revealed a 50-mm fusiform aneurysm in the right external artery, which extended to the right common femoral artery (**Figs. 1A** and **1B**). Following discussions with the cardiologist, we planned out a surgical graft replacement of the right iliofemoral aneurysm.

Using a retroperitoneal approach, the target aneurysm was resected and replaced by a 10-mm polyester graft (InterVascular, La Ciotat, France). The right deep femoral artery was left untouched due to its small size. Postoperative CT showed successful reconstruction of the right iliofemoral artery (**Fig. 2A**). However, 6 years later, the patient revisited our hospital for recurrence of right groin pain. CT revealed a 26-mm right deep femoral artery aneurysm (**Fig. 2B**). Reoperation was planned to rule out impending rupture, and distal reconstruction of the deep femoral artery was performed (8-mm Propaten, WL Gore, Flagstaff, AZ, USA) (**Fig. 2C**). The patient was discharged and did not encounter any further issues for the next 3 years. In addition to atherosclerotic changes, the vessel wall was thin and fragile in the first and second surgery. Histopathological investigation revealed tumor cells in the aneurysm wall, thereby confirming the diagnosis of vascular involvement in NF1 (**Figs. 3A** and **3B**).

Discussion

Although vascular involvement is uncommon in patients with NF1, aneurysm formation has been reported because of friable vascular tissue. Aneurysms can be formed in any vessel in the body, including the renal, intercostal, carotid–vertebral, and intracranial arteries.[1–4] However, an iliofemoral artery aneurysm in an NF1 patient is extremely rare.[5]

We considered two treatment options—surgical resec-

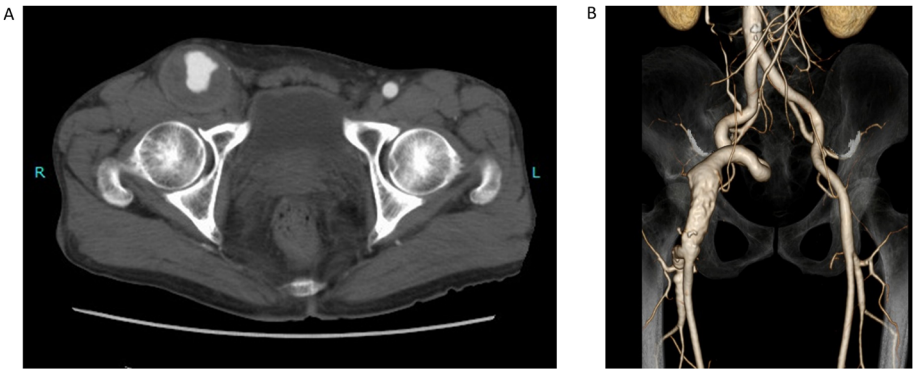

Fig. 1 Preoperative computed tomography showing a right iliofemoral aneurysm. (**A**) Axial image, (**B**) 3D image.

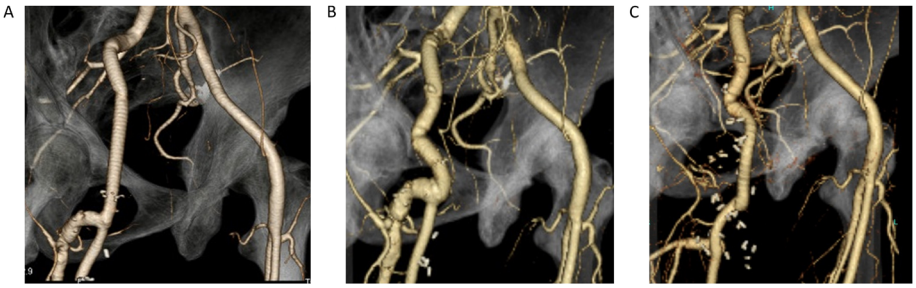

Fig. 2 Postoperative computed tomography images. (**A**) Reconstructed iliofemoral artery after the first surgery. (**B**) Progression of right deep femoral artery aneurysm after 6 years. (**C**) Reconstructed deep femoral artery after the second operation.

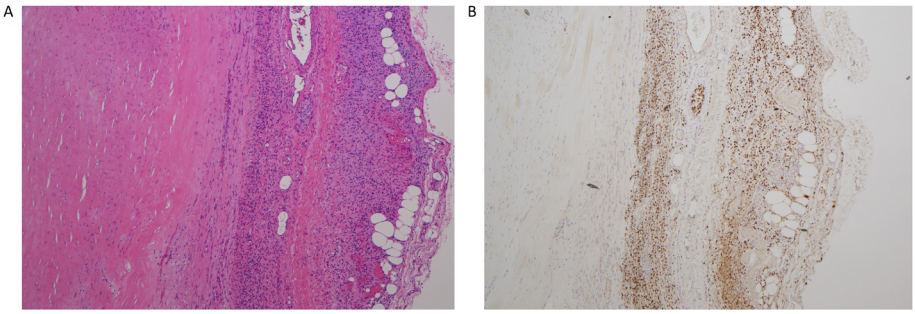

Fig. 3 Histopathology images. (**A**) Cross-section of the iliac aneurysm wall (hematoxylin–eosin, original magnification ×100). Zonal vascular smooth muscle cell proliferation, mesodermal dysplasia, proliferating wavy spindle cells are invading adventia, by invasion neurofibromatosis tissue. (**B**) With S100 immunoperoxidase staining of the aneurysm wall (original magnification ×100), proliferating cells were stained positive for S100 protein, indicating a neural origin.

tion and endovascular repair. Stent-graft insertion was not chosen because of the need to cover the deep femoral artery and the highly flexible area referred to as the nonstenting zone. Moreover, it is difficult to predict the long-term outcome of this disease due to its diffuse and progressive nature. However, endovascular treatment would have been a feasible option for a patient with a high surgical risk.

The right deep femoral artery was preserved in the first surgery because of its small size. It can be conjectured that the second surgery would not have been necessary if this segment had been resected in the first surgery. Nevertheless, we believe it resulted in an acceptable outcome based on the fact that this disease is progressive and can affect any part of the body. In our case, an additional aneurysm measuring 10 mm was detected in the splenic artery. Although this aneurysm has not grown in size over the course of 9 years, the aneurysm and the entire vascular

system require careful radiographic follow-ups as there is a higher probability of progression and/or rupture due to the fragile vascular tissue.[2,4,6]

The reason for groin pain was unclear. Compression of the surrounding structure is thought to be a cause of the pain. However, given the fragile vessel nature of this disease, it was difficult to rule out impending rupture despite the size of the deep femoral artery in the second operation.

Conclusion

We encountered a rare case of symptomatic iliofemoral aneurysm in a patient diagnosed with NF1. While surgical resections and constructions were successful, reoperation was ultimately required 6 years later. As the vascular tissue is thought to be fragile, careful radiographic follow-ups are necessary.

Acknowledgments

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Consent

The patient has given verbal consent for publication of this case report and any accompanying images and agreed to give written consent if required.

List of Abbreviations

CT: computed tomography

Disclosure Statement

All authors have no conflicts of interest.

Author Contributions

Manuscript preparation: TU
Critical review and revision: all authors
Final approval of the article: all authors
Accountability for all aspects of the work: all authors

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