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CASE REPORT

# Successful Surgical Reconstruction of a Ruptured Brachial Artery Aneurysm in a Patient With Type 1 Neurofibromatosis

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**WHAT UNIQUE EDUCATIONAL MESSAGE IS PROVIDED AND WHY IS IT RELEVANT?**
This case describes the successful surgical reconstruction of a challenging vascular pathology in a patient with type 1 neurofibromatosis. The article is intended to provide tools for future successful vascular reconstruction of a ruptured brachial artery aneurysm.

**Introduction:** The vascular pathology of patients with type 1 neurofibromatosis (NF-1) is known. Aneurysms of the brachial artery in NF-1 patients are rare and surgical treatment remains a challenge.
**Report:** A patient known to have NF-1 presented with swelling of the left arm. Computed tomography angiography showed a ruptured aneurysm of the brachial artery. Operative reconstruction was performed using reversed saphenous vein.
**Discussion:** Up to now four cases had been published describing brachial aneurysms in NF-1 patients. This case describes the successful reconstruction of a ruptured brachial aneurysm, using a saphenous vein.

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## INTRODUCTION

The case of a 48 year old woman with a history of type 1 neurofibromatosis (NF-1) is described. She presented to the emergency department with progressive and painful swelling of the left upper arm. Investigations showed a rare ruptured brachial artery aneurysm.

## CASE REPORT

A 48 year old woman presented to the emergency department because of progressive and painful swelling of the left upper arm. She was known to have NF-1. There was no history of trauma or infection. Her previous medical history included a spontaneous haemothorax caused by a neurofibroma and removal of a pheochromocytoma.

She was not acutely ill, apyrexial (37.4°C), and had a swelling of the medial side of the left upper arm. There was no loss of sensation and no wrist drop.

During her stay in the emergency room, the patient remained haemodynamically stable Blood tests showed a haemoglobin of 8.4 g/dL and no increase in infection parameters (leucocytes 6.3 × $10^9$/L, C reactive protein 14 mg/L).

Ultrasonography of the left upper arm revealed two masses along the left brachial artery with an arterial flow pattern. Computed tomography angiography (CTA) was performed and three true aneurysms were detected. Two large broad based aneurysms were located in the left brachial artery (respectively 24 × 14 mm and 22 × 12 mm), with a small aneurysm between (10 mm). There was active extravasation of contrast from the small aneurysm causing a large soft tissue haematoma (Fig. 1A–C). CTA of chest, abdomen, and upper legs demonstrated no further aneurysms.

Direct surgical intervention was indicated. An incision was made on the medial side of the upper arm. The brachial artery including the aneurysms was resected over a length of 10 cm. The vascular tissue appeared to be fragile. Reconstruction was performed using reversed saphenous vein to create a bypass with end to end anastomosis. Peroperatively, the reconstruction appeared successful, confirmed by Doppler signals and palpable radial pulse.

Post-operatively the patient remained stable, the pain decreased, and she recovered without complications. After four weeks of follow up, the patient mentioned a persisting decrease in strength and sensation in the left hand.

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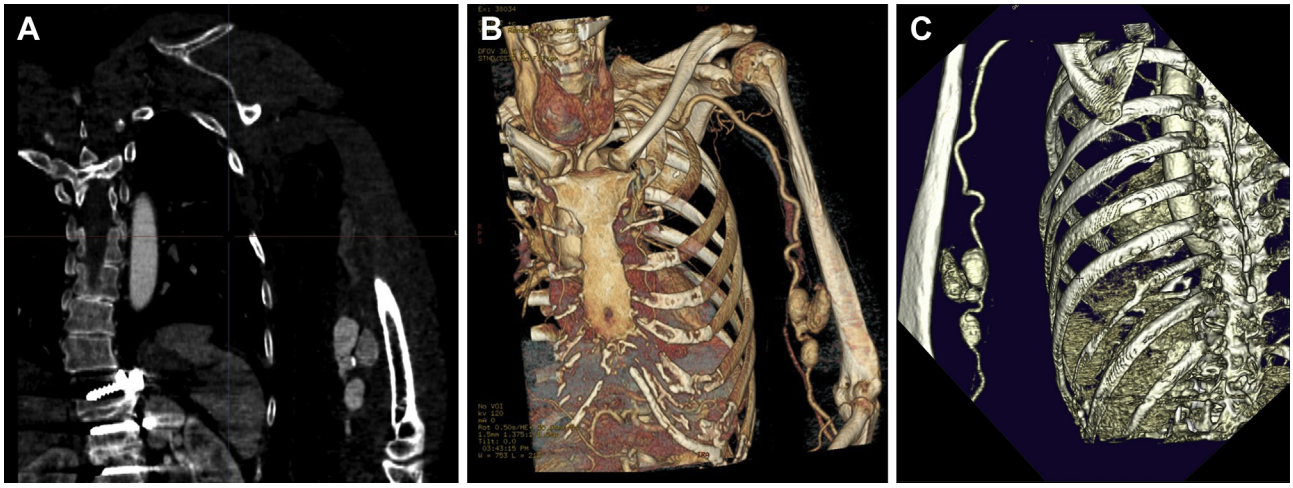


**Figure 1.** Computed tomography angiography (CTA) and three dimensional images of the three aneurysms. CTA of the left upper arm showed a ruptured aneurysm originating from the brachial artery. Axial (A) view of a ruptured aneurysm with contrast extravasation and soft tissue haematoma. Three dimensional volume rendered images of three aneurysms of which the middle, small aneurysm is ruptured. (B) Anterior view. (C) Posterior view.

## DISCUSSION

Although aneurysmal vascular disease is a known complication in patients with NF-1, literature about the pathology and treatment of choice is scarce. Aneurysms of the brachial artery in patients with NF-1 are rare, with only four case reports being published between 1998 and 2010. Among these four cases, two patients died after surgery and in only one case was a successful repair performed.[1—4] The condition should not be underestimated since it has been shown to be life threatening.

Neurofibromatosis, an autosomal dominant genetic disorder, affects one in 400—2600 individuals.[5—7] It is also known as Von Recklingshausen disease and mostly affects the brain, spinal cord, skin, eyes, and long bones. NF-1 is characterised by the occurrence of cutaneous café au lait spots, formation of benign neurofibromas, and iris hamartomas.[8]

Vascular pathology is a complication of NF-1, affecting 0.4—6.4% of patients.[9] The most frequently reported vascular anomalies are intracranial aneurysms, stenosis, and arteriovenous malformations.[9] The most common sites for aneurysms are the aorta, visceral arteries (including hepatic, superior mesenteric, gastroduodenal, and renal arteries), cerebrovascular and vertebral arteries.[6,9]

Aneurysms are mostly asymptomatic and are often discovered when they rupture.[6,9] Involvement of peripheral arteries is uncommon, with clinical manifestations including acute swelling, haematoma, pain, and possible hypovolaemic shock in rupture cases.[3]

The vascular manifestation of NF-1 is described as smooth muscle cell loss and proliferation of the intima with fibrous thickening, contributing to neurovascular malformations in smaller vessels.[10] In larger vessels, direct tumour invasion causes tissue compression of the vasa vasorum and wall weakening.[4] Because neurofibromatosis causes weakening of the vessels and surrounding tissue, surgical reconstruction is a challenge.

Among the four previously reported cases, two describe resection of the artery, with one patient dying post-operatively.[3] One article describes a failed saphenous vein reconstruction complicated by post-operative haemorrhage, resulting in amputation of the arm.[1] The only known successful reconstruction to date is described by Emori et al.,[4] using the great saphenous vein for interposition.

There is a lack of consensus whether routine screening for vascular lesions is indicated in all patients with NF-1.[4] The patient was assessed post-operatively but no other vascular aneurysms were found on imaging.

Up to now, four cases had been published describing brachial aneurysms in a NF-1 patient. This is the second case describing successful reconstruction of a ruptured brachial aneurysm using a saphenous vein.

## CONFLICT OF INTEREST

None.

## FUNDING

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