Ruptured aneurysm of the ulnar artery in a woman with neurofibromatosis

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A 61-year-old woman with neurofibromatosis type 1 (Recklinghausen's disease) was referred for massive swelling of the right forearm, pain, increasing numbness, and impaired movement of the fingers. Angiography demonstrated a 13- × 11-mm aneurysm and a capped rupture of the ulnar artery. Because of the complicated soft-tissue condition, interventional treatment was indicated. Two 360° coils were placed for embolization of the ruptured aneurysm. Arterial involvement in neurofibromatosis is a well known but infrequent occurrence. Stenotic lesions predominate. Aneurysmal defects are less common, and rupture of peripheral arteries is exceptional. (J Vasc Surg 2009;49:494-6.)

Aneurysms of the ulnar artery are very rare. Systematic descriptions in the literature may be almost exclusively found within the context of the hypothenar-hammer syndrome. Neurofibromatosis is an autosomal-dominantly inheritable neuroectodermal tissue disorder. Depending on the gene location, type I (chromosome 17q11.2) may be distinguished from type II (22q12). Type I presents with typical café-au-lait macules and neurofibromas. Characteristics for the rarer type II are neurofibromas as well as schwannomas of the vestibular nerve and a juvenile subcapsular cataract.

Vascular pathologies in connection with neurofibromatosis type I have been systematically described by Reubi.[1] They include renovascular hypertension, aortic stenoses, occlusive arterial disease, and aneurysms of the pulmonary, cardiac, cervical, and cerebral arteries. However, the main locations for the stenoses and aneurysms are the aortal branches and the abdominal aorta.[2] The literature documents a number of real and pseudoaneurysms at peripheral sites.[2-7] To our knowledge, no authors have reported an aneurysm of the ulnar artery within this context. We report the case of a 61-year-old woman with a ruptured ulnar aneurysm.

CASE REPORT

The patient presented to the emergency department with a painful, spontaneous marked acute swelling of the right forearm and an increasing numbness and impaired movement of the fingers. The patient had no history of acute or remote trauma. The ulnar pulse could not be palpated, but there was a weak radial pulse and a normal brachial pulse.

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Competition of interest: none.

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The patient had been diagnosed in childhood with neurofibromatosis type I. She underwent two operations, in 1994 and 1995, on the right hand for the removal of neurofibromas (arthrodesis with a mini-plating for metacarpal V dislocation). During the first operation, the ulnar artery was ligated distally to the wrist because it was encapsulated by the tumor. The main tumor mass of the removed neurofibromas was located in the area of the hypothenar and thenar.

Apart from hypertension, she had no other medical conditions and no major impairments in every-day-life as a result of the disease. Ultrasound imaging showed a normal flow curve of the radial artery as well as a major hematoma of the medial forearm. Angiography showed normal images of the axillary artery and the brachial artery, including their branches. The radial artery presented as the dominant vessel in the forearm. The ulnar artery was not to be distinguished in its normal course; here, a thin meandering vessel filled at the level of the enlarged soft tissue with imaging of an aneurysm of 13- × 11-mm size and a capped rupture with a contrast medium cavity of 19- × 15-mm size (Fig 1).

This finding was approximately 7 cm proximal to the wrist. Because of the complicated soft-tissue condition, interventional treatment was indicated. A 5F sheath was introduced through the brachial artery, and a 2.3 Tracker catheter (Cordis/Johnson & Johnson, Miami, Fla) was advanced and superselectively placed at the aneurysm. Two 360° Guglielmi detachable coils (GDC, Target Therapeutics/Boston Scientific, Fremont, Calif) were placed (14/30 and 10/30). Occlusion of the aneurysm was confirmed after the second coil was placed (Fig 2). Angiography showed no retrograde filling of the aneurysm through the radial artery or the palmar arch. The sheath was removed at the end of the procedure.

After the embolization, we applied a continuous intravenous (IV) heparin infusion and iloprost (40-μg IV) for 24 hours. Analgesia, antiphlogistic treatment, and cold application were supportive. A few hours after the intervention, the patient reported a marked decrease in pain and a complete recovery of the sensitive and motor impairments was observed. The patient was discharged home on the third day after the intervention without any discomfort (Fig 3). A complete screening of the vascular tree by magnetic resonance angiography 3 months after the intervention showed no additional aneurysms.

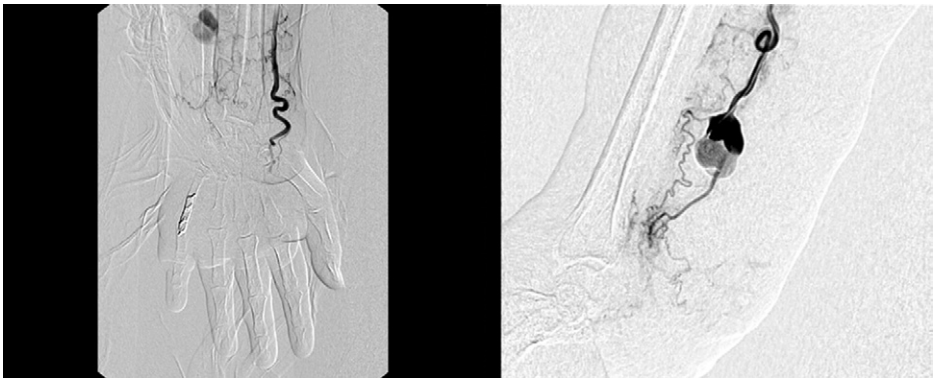

Fig 1. A, The diagnostic angiography revealed a 13- × 11-mm aneurysm and **(B)** a capped rupture with a 19- × 15-mm contrast medium cavity approximately 7 cm proximal to the wrist.

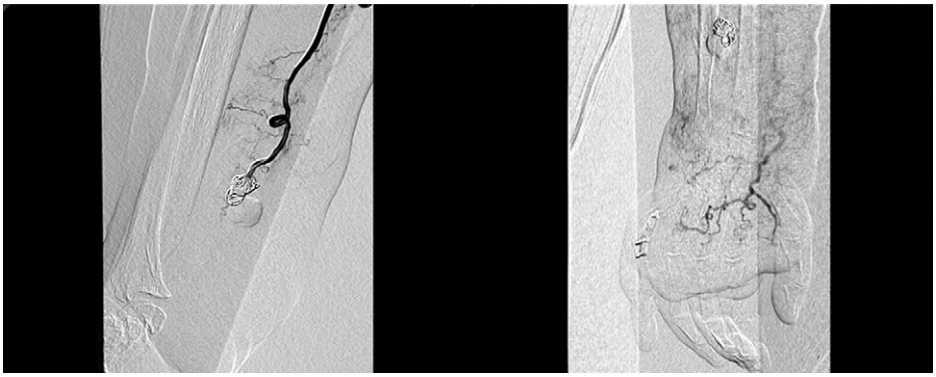

Fig 2. A, Occlusion of the aneurysm through placement of two 360° GDC coils. **B,** After placement of the second coil, the aneurysm was confirmed to be occluded. The radial artery presented as the dominant vessel in the forearm.

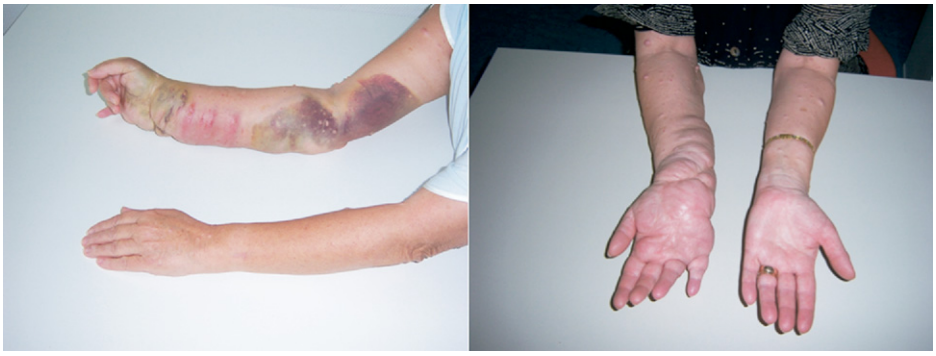

Fig 3. Macroscopic findings on the **(A)** second postinterventional day.

DISCUSSION

The case presented here is to our knowledge the only one that has been described in association with neurofibromatosis type I. The association between neurofibromatosis type I and stenotic vascular alterations is well documented in the literature, but aneurysms of the peripheral arteries are very rare. Four histopathologic characteristics have been described for neurofibromatosis-associated vascular stenoses: pure intimal, advanced intimal, intimal aneurysmal, and nodular.[1,5,8] At the expense of the muscular and elastic parts of the vascular wall, there is an intimal hyperplasia and a nodular thickening of the adventitia. Feyrter[9] described an epithelioid form in which the whole vessel is infiltrated with neural cells. In 1974 Salyer[10] postulated that the proliferation of Schwann cells was the common pathogenetic basis, a theory that later studies have supported.[11] On

the grounds of the vessel size, Greene et al[12] distinguished two patterns of damage: (1) direct infiltration of the vascular wall by Schwann cells with intima thickening and (2) destruction of the media and the elastic fibers that may lead in larger vessels to the formation of a stenosis or an aneurysm. In small-bore vessels a mesodermal dysplasia dominates. This leads to stenoses and only occasionally to a poststenotic aneurysm.[12] The histomorphologic picture resembles generally that of polyarteritis nodosa; whereas in contrast to this, perivascular inflammation and adjacent tissue necrosis are missing.[1,3] Clinically and experimentally there are hints for a tumor angiogenesis factor that triggers the endothelial proliferation and vasodilation.

A general screening is recommended in Ehlers-Danlos and Marfan syndrome, osteogenesis imperfecta, pseudoxanthoma elasticum, and neurofibromatosis to prevent fatal outcomes of occult aneurysms.[4,13] Although the incidence of the disease is relatively high (40:100,000), vascular involvement is rather rare (<3%); therefore, we consider routine screening not to be feasible. Patients with type I or type II neurofibromatosis and hypertension, those with neurofibromas along the peripheral vascular axes, and symptomatic patients as well as those who have been treated for a vascular incidence, should be screened generally. We consider whole-body magnetic resonance angiography, including the cerebral vasculature, at 3- to 5-year intervals to be the most appropriate screening method. All aneurysms >1 to 1.5 cm in diameter should be treated.

We chose an interventional treatment for our patient because of the difficult soft-tissue conditions with a history of several operations on the forearm, a dominant radial artery, and a proper deep palmar arch. Despite the developing neurologic symptoms, we chose this procedure because we considered an open intervention to be an additional trauma.

The clinical signs did not indicate a manifest compartment syndrome. If a compartment syndrome had developed in the course of treatment, we would have opted for secondary splitting of the compartment. In this case we would have relied on aggravating clinical symptoms rather than the other option used at our department of measuring the compartment pressure with a cannula. In alert patients we pay particular attention to the medical history and clinical evaluation. In comatose patients we would perform a compartment splitting at a compartment pressure of ≥20 mm Hg. In our patient a decompression of the area would have been necessary, anyway, in case of nonreceding clinical symptoms. Fortunately, the symptoms were actually improving rapidly. We ascribe this to the patient having presented in an acute situation after the rupture. Pressure relief only and prevention of further extension have facilitated the regression of neurologic symptoms.

As did other authors with a similar case,[3] we asked ourselves whether direct iatrogenic arterial damage from the previous operations might have caused the aneurysm. However, we consider this assumption to be unlikely in light of the long time (12 years) that had passed. According to the surgical records, no previous manipulations had been done near the site of the (later) aneurysm. The scar ends at a distance of >5 cm from the aneurysm. The presence of neurofibromas requiring surgical intervention also suggests disease activity in the area of the aneurysm; therefore, we consider the neurofibromatosis as such to be the most likely cause.

Because of the endovascular intervention and the situation of single-vessel perfusion of the hand, we heparinized the patient for 24 hours. This is standard procedure at our department. Because of the severe clinical symptoms before the intervention, iloprost was given short-term to improve the microcirculation.

In conclusion, taking into account our own experience, we consider a screening for further aneurysms to be sensible in symptomatic patients with Recklinghausen's disease and those with hypertension or neurofibromas in relevant vascular regions. The interventional approaches have proven their value, especially in vascular regions that are difficult to access or may be accessed only with a major surgical trauma.[6] In our patient we were able to obtain a very favorable treatment outcome with a maximum of functionality by means of a minimally invasive approach.

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