

## SHORT REPORT

## Endovascular Treatment for a Ruptured Lumbar Artery Aneurysm in a Patient with von Recklinghausen Disease

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**Introduction:** Vasculopathy, such as an aneurysm, stenosis, rupture, or arteriovenous fistula, in patients with neurofibromatosis type I (NF-1; von Recklinghausen disease) is well recognised. However, there has been no report regarding treatment for a ruptured lumbar artery aneurysm associated with NF-1. We present the first report of successful endovascular treatment by coil embolisation for a ruptured lumbar artery aneurysm in a patient with NF-1.

**Report:** A 52 year old man with a history of NF-1 was referred with back pain and anaemia. The computed tomography scan showed rupture of a solitary lumbar artery aneurysm. The rupture was successfully treated by endovascular embolisation with a coil and *N*-butyl-2-cyanoacrylate.

**Discussion:** Endovascular treatment with coil embolisation was performed safely in this patient.

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### INTRODUCTION

Neurofibromatosis type 1 (NF-1; von Recklinghausen disease) is an autosomal neuro-cutaneous disorder with a prevalence of about 1 in 3,000 births; it is caused by a mutation of *NF1* located on the long arm of chromosome 17. The typical clinical manifestations are café-au-lait macules, freckling, peripheral neurofibromas, Lisch nodules, and bone abnormalities. Vasculopathy, such as an aneurysm, stenosis, rupture, or arteriovenous fistula, in patients with NF-1 is well recognised, but is rarely encountered clinically. Treatment of peripheral artery aneurysms, such as in the internal carotid artery, renal artery, subclavian artery, or intercostal artery, has been reported,<sup>1–4</sup> but there has been no report of an aneurysm in the lumbar artery of a patient with NF-1. The first case of successful endovascular treatment with coil embolisation for a ruptured lumbar artery aneurysm in a patient with NF-1 is described.

### CASE REPORT

A 52 year old man with a history of NF-1 was referred for back pain that continued for 2 days. The computed tomography (CT) scan showed a large haematoma in the right

retroperitoneal space due to rupture of a right lumbar artery aneurysm (Fig. 1). Clinical examination showed multiple café-au-lait macules and right flank pain. His vital signs were stable, but the blood test results indicated anaemia (hemoglobin level 8.2 g/dL).

The emergency angiogram through the right femoral artery showed active extravasation of contrast medium from the ruptured aneurysm of the third right lumbar artery, which was successfully selected using a manually curved 4 Fr hook shaped catheter (RC-09; Medikit, Tokyo, Japan) (Fig. 2). At the distal side of the aneurysm, the lumbar artery branched into three. Through the 4 Fr catheter, a 2.9 Fr catheter (microcatheter with two coils for support; Hi-Lex, Takarazuka, Japan) was coaxially inserted using a 0.018 inch guidewire (AQUA<sup>®</sup>VIII guidewire; Cardinal Health, Dublin, OH, USA). Three distal arterioles were successfully embolised using several microcoils (Tornado<sup>®</sup> Embolisation Coil, MReye<sup>®</sup> Embolisation Coil; Cook Medical, Bloomington, IN, USA), measuring 2–5 mm in diameter (Fig. 3). The aneurysm and residual tiny arteriole originating from the aneurysm were embolised using a mixture of *N*-butyl-2-cyanoacrylate and Lipiodol (Guerbet, Paris, France) in a 1:2 ratio. The inflow artery was embolised using the same microcoils measuring 2–4 mm in diameter. The final angiogram showed that the third right lumbar artery aneurysm was embolised successfully (Fig. 4). No blood transfusion was required and no complication was observed. The post-operative enhanced CT scan showed a thrombosed lumbar artery aneurysm with no extravasation

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**Figure 1.** Computed tomography (CT) scan of the large haematoma. CT revealed a large haematoma in the right retroperitoneal space due to rupture of the right lumbar artery aneurysm.



**Figure 2.** Emergency angiogram through the right femoral artery. The arrow shows active extravasation of contrast from the ruptured third right lumbar artery aneurysm.

and no additional aneurysm. The brain magnetic resonance imaging scan showed no aneurysm in the cervical or carotid arteries. The patient was discharged home in a good condition.

## DISCUSSION

The reported prevalence of vascular abnormalities in patients with NF-1 is 0.4–6.4%.<sup>5–7</sup> As most patients with vascular abnormalities are asymptomatic, the rate may be underestimated. Past studies have shown that renal artery stenosis is most frequently observed, and abdominal aortic coarctation, internal carotid artery aneurysms, and cervical vertebral arteriovenous malformations are other common manifestations. A lumbar artery aneurysm has been reported only once, and the patient was observed



**Figure 3.** Digital subtraction angiogram after embolisation with a coil and *N*-butyl-2-cyanoacrylate. The outflow artery is shown.



**Figure 4.** Digital subtraction angiogram after embolisation of the third right lumbar artery. There is no contrast in the aneurysm.

conservatively.<sup>1</sup> As far as the authors are aware the present case is the first report of endovascular treatment for a ruptured lumbar artery aneurysm associated with neurofibromatosis.

Oderich et al. reported 41 vascular abnormalities repaired in 23 patients with NF-1, reconstructions in 15 and endovascular treatment.<sup>1</sup> Freedom from graft related complications was 83% at 10 years. Oderich et al. concluded that operative treatment of symptomatic patients with NF-1 was safe, effective, and long lasting.

Endovascular treatment has replaced open surgery as it is less invasive and surgical repair of a fragile artery in patients with neurofibromatosis is difficult.<sup>2</sup> In the present patient, coil embolisation was selected because it may be difficult to

detect and ligate the point of rupture inside the haematoma.<sup>3</sup> When the endovascular approach is considered in a patient with NF-1, attention should be paid to avoid injury to the friable vascular endothelium.<sup>4</sup> Moreover, physicians have to examine the entire body as there may be multiple vascular lesions. In general, survival is shorter in patients with NF-1 than in the general population.<sup>8</sup> The most common cause of death is malignancy, followed by vascular disease, especially in younger patients. Close observation during the follow-up period is also mandatory in patients with NF-1.

#### CONFLICT OF INTEREST

None.

#### FUNDING

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