Endovascular treatment of contained rupture of a superior mesenteric artery aneurysm resulting from neurofibromatosis type I

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A 31-year-old woman with neurofibromatosis type I (NF-I) came to our hospital with hypotension and abdominal pain. A computed tomography (CT) scan showed blood in the retroperitoneum and two saccular aneurysms in the superior mesenteric artery (SMA). The largest measured 2.5 cm in diameter. She was treated with placement of a covered stent in the SMA, and both aneurysms were excluded from the systemic circulation. Arterial aneurysms are rare in this disease, and rupture of an SMA aneurysm in this context had been reported only once. We report an unusual case of a contained rupture of an SMA aneurysm associated with NF-I, successfully treated with a covered stent. (J Vasc Surg 2010;51:461-4.)

Neurofibromatosis type I (NF-I), or von Recklinghausen disease, is an autosomal dominant disorder affecting one in 3000 individuals.[1] Cardinal features of NF-I include multiple café-au-lait macules, benign neurofibromas, and iris hamartomas.[1] Arterial involvement has also been noted, usually in the form of stenoses. Aneurysms involving the superior mesenteric artery (SMA) have been infrequently reported, and rupture of an SMA aneurysm in this context had been reported only once.[2] We submit an unusual case of a contained rupture of an SMA aneurysm in a patient with NF-I, treated successfully with a covered stent.

CASE REPORT

A 31-year-old woman came to the emergency department of our University Hospital with hypotension (90/60 mm Hg) and abdominal pain. She had these symptoms for approximately 6 hours. The patient had been diagnosed in childhood with NF-I when a benign neurofibroma was resected from her abdomen at age 6. She had multiple café-au-lait skin macules and subcutaneous neurofibromas in both legs. She had no history of diabetes mellitus, hypertension, or tobacco use. She denied any history of previous abdominal trauma or intravenous drug abuse. She was not taking any medications.

The emergency department doctor administered intravenous fluids, and her blood pressure increased to 110/70 mm Hg; the patient was alert, and her heart rate was 82 beats/minute. An emergency computed tomography (CT) scan showed blood in the retroperitoneum (Fig 1) and two saccular aneurysms arising from the SMA (Fig 2), without active bleeding. The largest measured 25 mm in diameter, and the second aneurysm measured 17 mm.

The patient was taken immediately to the angiography suite. The right common femoral artery was punctured, and the SMA was selectively catheterized with a 5F internal mammary catheter. At this point, 3500 units of intravenous heparin were administered. Angiography confirmed the presence of two saccular aneurysms arising from the SMA (Fig 3), with no active bleeding at the moment. The catheter was negotiated over a 0.035-inch guidewire into the distal portion of the vessel. At that point, a 0.035-inch stiff Rosen wire was advanced through the catheter, and the catheter was removed. An 8F catheter sheath had been placed in the right groin over the Rosen wire. Through the sheath, a 7F guiding catheter was placed over the Rosen wire in the proximal portion of the SMA. A polytetrafluoroethylene (PTFE)-covered stent (Advanta V12, 6 × 38 mm, Atrium Medical Corporation, Hudson, NH) was advanced over the guidewire into the SMA. Arteriography was then performed to find the best position for the covered stent, and it was then inflated to 6 mm. After its placement, an arteriography showed that both aneurysms were excluded from the systemic circulation (Fig 4). At least three jejunal branches were covered by the stent, but the patient did not have any symptoms of bowel ischemia, because the collateral circulation was very good. A follow-up CT scan (1 month after the procedure) showed patency of the SMA, correct position of the stent, and confirmed the exclusion of both aneurysms from the systemic circulation (Fig 5).

The patient made a full recovery, with no major adverse events and was discharged 2 days postoperatively. At a 6-month review, the duplex B-mode images did not show any turbulence or stenosis proximal, intra, or distal to the covered stent. We did not see any leaks (there was no flow inside the aneurysms). The fasting duplex ultrasound scan peak systolic velocities were 184 cm/second in the SMA proximal to the stent, 191 cm/second intra-stent, and 191 cm/second in the SMA distal to the stent.

DISCUSSION

Neurofibromatosis was first described by von Recklinghausen in 1882.[3] It is an autosomal dominant inheritable

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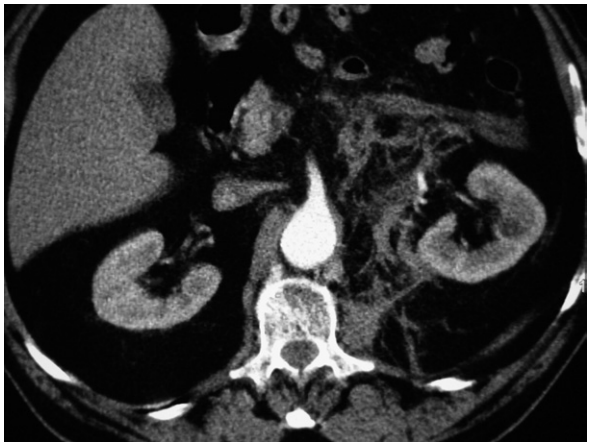

Fig 1. Computed tomography (CT) scan showing blood in the retroperitoneum.

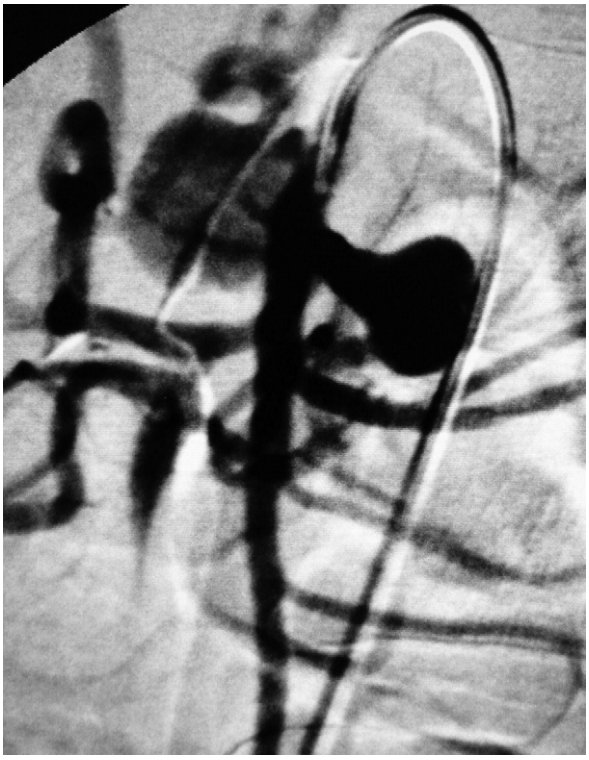

Fig 3. Selective angiography of the superior mesenteric artery (SMA), demonstrating two saccular aneurysms in this vessel, before the implant of a covered stent.

Fig 2. Computed tomography (CT) scan showing two saccular aneurysms arising from the superior mesenteric artery (SMA).

neuroectodermal tissue disorder. Heterogeneity is the clinical hallmark of neurofibromatosis, and at least seven varieties of the disease have been described, of which type I is the most common.[4]

NF-I, also called von Recklinghausen disease, presents with typical café-au-lait macules, benign neurofibromas, and iris hamartomas.[1,5] It has been shown that the gene mutation in NF-I is located in the pericentric region of chromosome 17.[6-9]

The prognosis for neurofibromatosis, which is unpredictable for a given subject, depends essentially on the existence of cerebral tumors, which are responsible for death in 72% of cases.[10]

Vascular involvement in NF-I has been systematically described by Reubi[11] in 1944. He described three forms of vascular lesions, depending on the size of the vessel: pure intimal, intimal-aneurysmal, and adventitial-nodular. He noted intimal proliferation with breakdown of muscle and elastic layers and adventitial nodular thickening. Feyrter[12] subsequently described an epitheloid form, with involvement of the entire vessel wall by neural cells. In 1974, Salyer and Salyer[13] proposed Schwann cell proliferation as the common pathogenesis. That same year, Greene et al[14] defined two different lesions based on vessel size. In larger vessels, there was direct invasion by Schwann cells and intimal thickening and destruction of the media and elastic tissue, leading to either stenosis or aneurysm. In smaller vessels, mesodermal dysplasia caused a proliferation of smooth muscle in the intima resulting in stenotic lesions and occasionally poststenotic aneurysms.

Arterial stenosis has been the most commonly reported lesion. Cases of renal artery stenosis have been the most frequent.[14,15] Celiac, mesenteric, iliac, and intracranial artery stenoses have also been noted. Arterial aneurysms have been less commonly described. These have often been

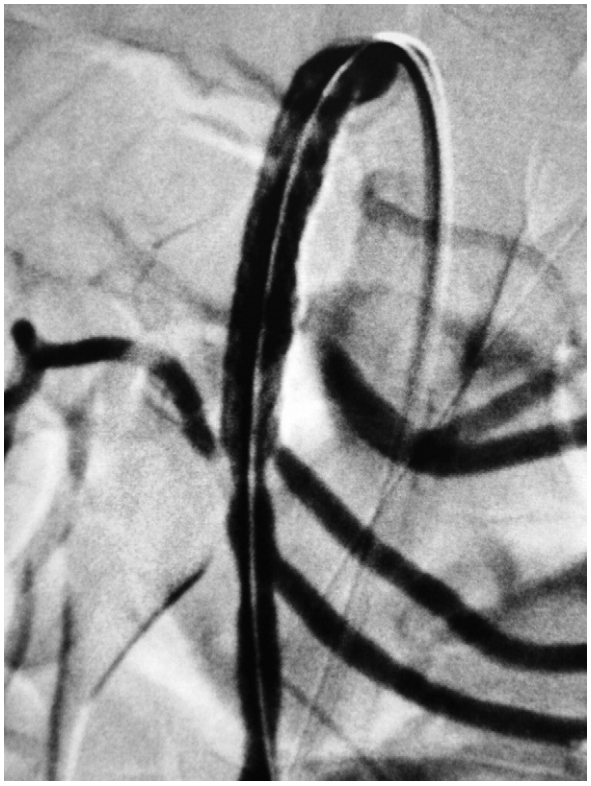

Fig 4. Selective angiography of the superior mesenteric artery (SMA) after the implant of a covered stent: both aneurysms were excluded from the systemic circulation.

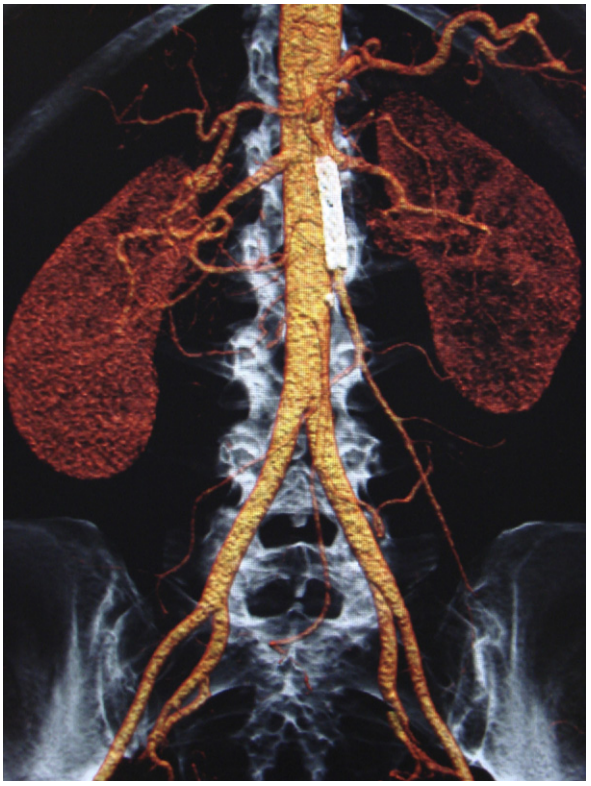

Fig 5. Follow-up computed tomography (CT) scan (1 month after the procedure) showing patency of the superior mesenteric artery (SMA), correct position of the stent, and exclusion of both aneurysms from the systemic circulation.

associated with poststenotic dilatations. Again, renal artery involvement seems to predominate.[2]

Aneurysms of the visceral arteries are exceptional in this disease; the major concern is the splenic artery.[16] Only eight cases of SMA aneurysms have been reported in NF-I.[2,15-19] Zochodne[15] reported a 2-cm diameter aneurysm of the SMA associated with coarctation of the abdominal aorta and poststenotic changes in the renal arteries. Fye et al[16] described multiple small aneurysms on the branches of the SMA. Henley and Kaude[17] mentioned an SMA associated with multiple renal artery aneurysms but did not detail the case. Hassen-Khodja et al[18] reported a case with two aneurysmal defects: one on the common hepatic artery (treated with embolization) and another on the SMA (not treated). Huffman et al[2] reported a patient who underwent ligation of a ruptured SMA aneurysm. Finally, Cormier and Cormier[19] reported three cases: one involving only the SMA and the two others associated with thoracoabdominal coarctation. Our patient had two aneurysmal defects on the SMA, presented with contained rupture, and underwent successful endovascular repair.

The histopathologic features of arterial lesions in NF-I disease consist of fibrodysplastic changes in the media with segmental fragmentation of the vessel wall caused by atrophy of the muscularis and formation of saccular aneurysms. Both aneurysms in our patient were of the saccular type. Fusiform aneurysms are less common and are often poststenotic.[11,13,14]

The pathophysiologic characteristics of vascular lesions in neurofibromatosis remain controversial. The alterations observed in the media and the adventitia appear compatible with the mesodermic and neuroectodermic tissue involvement in this disease.[5,11,13,14]

The silent nature of most vascular lesions and the inaccessibility of involved vessels to clinical examination have resulted in an underappreciation of its occurrence. An autopsy series of NF-I patients who died from other causes found vascular abnormalities in eight (44%) of 18 cases.[13] Screening of all patients with NF-I for arterial disease should be performed. Noninvasive imaging modalities such as duplex ultrasound scanning, magnetic resonance angiography, and CT scanning are useful means of visualizing vessels. Reports of involvement of almost every major artery can be found in the literature, and no vessels are spared, including veins.[20]

We decided to offer the endovascular treatment to our patient because the origin of both aneurysms in the SMA were very close, and a covered stent could exclude both aneurysms from the systemic circulation at the same time. In addition, the patient had a previous abdominal operation at age 6 to remove a neurofibroma very close to the

SMA, and we thought that another abdominal operation should be avoided. Given the patient's young age and the long-term concerns of SMA stent stenosis/occlusion, we recommend follow-up with duplex ultrasound scan at a 6-month interval and continuous use of aspirin and clopidogrel. If a significant stenosis is found, the SMA stent should undergo percutaneous transluminal angioplasty to avoid occlusion and potential bowel ischemia. Because histologic confirmation of the nature of the SMA aneurysms was not obtained, their relationship to the patient's neurofibromatosis remains unproven, but it seems most probable. To our knowledge, this is the first patient who underwent successful endovascular repair of a contained rupture of an SMA aneurysm resulting from NF-I.

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