Late conversion after endovascular abdominal aortic aneurysm repair in a patient with Ehlers-Danlos syndrome

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ABSTRACT

Vascular Ehlers-Danlos syndrome is associated with life-threatening events. The management of the disease is challenging because of the emergency presentation of symptoms and the tissue friability of the aorta. We describe the successful treatment of a late type I endoleak after previous EVAR. (J Vasc Surg Cases and Innovative Techniques 2019;5:1-3.)

Keywords: Ehlers-Danlos syndrome; Endovascular repair; Open repair; Endoleak; Connective tissue disorders

Ehlers-Danlos syndrome (EDS) encompasses a group of rare genetic connective tissue disorders (CTDs). The vascular type (type IV) constitutes <5% of all cases but is associated with life-threatening events and high mortality rate because of the risk of vascular vessels' degeneration and rupture.[1,2] Thoracic endovascular aortic repair or endovascular aneurysm repair (EVAR) for patients with CTDs can be associated with late endoleaks in the sealing zones as a result of progressive degeneration of the aorta[3,4]; there is only one previous report of a patient with EDS and abdominal aortic aneurysm who was treated by EVAR.[5] Herein, we describe the long-term outcomes in a patient with EDS who was admitted to our department for severe abdominal pain and evidence of progressive type IA and IB endoleaks 7 years after EVAR. Informed consent for publication of this case report was obtained.

CASE REPORT

A 76-year-old man with abdominal and back pain presented to our hospital. His past medical history revealed spontaneous perforation of the sigmoid colon at a young age and also aortic arch and valve replacement at the age of 50 years. After the episode of hollow organ perforation, molecular genetic testing showed that the patient had EDS type IV. He had a history of EDS in his family with evidence of this connective tissue disease in four of the overall six siblings. In addition, he had a history of atrial fibrillation, arterial hypertension, hyperlipidemia, and hypothyroidism. Seven years ago, he had undergone endovascular repair of an abdominal aortic aneurysm 6 cm in diameter with placement of an aortobi-iliac endograft with suprarenal fixation at another hospital. Computed tomography angiography at the time of his admission to our department revealed a type IA and type IB endoleak (Figs 1 and 2) with a 7.3-cm-diameter aneurysm sac expansion (Fig 3).

The patient was immediately transferred to the operating room. A midline incision was made and the bowel was moved laterally. To prevent unexpected bleeding due to the fragility of the aorta, the infradiaphragmatic space was prepared immediately. A cautious proximal control was achieved by cross-clamping both the aorta and the endograft main body just below the renal arteries because the left renal vein was divided. After proximal and distal clamping and saccotomy, migration and malapposition of the graft in the dilated neck, and bleeding between the wall of the posterior iliac arteries and the limbs of the endograft, the presence of the type IA and type IB endoleaks was confirmed. The stent was explanted from the proximal aortic section down to the common iliac arteries. A polyester graft (18- × 9- × 9-mm in diameter) was then anastomosed with the end-to-end technique to the proximal aorta, to the right common iliac artery, and to the external iliac artery on the left. The sac was closed with the inlay technique. The abdomen was closed in layers. The patient had an uneventful recovery and was discharged to home on postoperative day 11. One year after the procedure, computed tomography angiography revealed good patency of the aortobi-iliac graft reconstruction and absence of de novo or para-anastomotic aneurysms (Fig 4).

DISCUSSION

CTDs represent a rare hereditary cause for the development of aortic aneurysms and dissections. The vascular type of EDS has an estimated prevalence of 1:100,000 to 1:250,000 births.[6] Vascular manifestations are among the most severe complications and involve various arterial and venous anomalies.[1] Although it is almost certain that the patients will require vascular intervention at some point in their lifetime, the optimal treatment is still under debate.

In the presented case, the patient suffered from abdominal and back pain due to the presence of type IA and type IB endoleaks 7 years after EVAR. The absence

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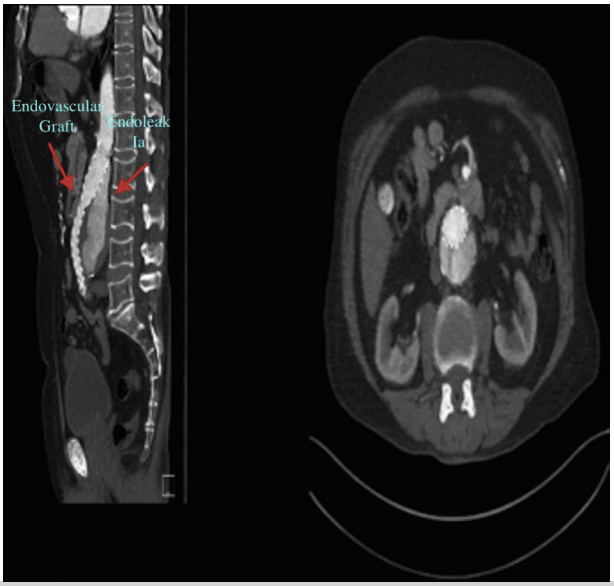

Fig 1. Type IA endoleak (sagittal and axial views).

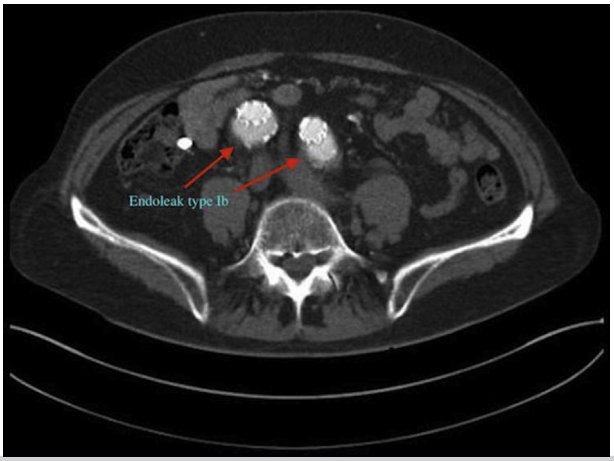

Fig 2. Type IB endoleak (axial view).

of an adequate seal zone determines the available urgent treatment options, including chimney EVAR and the open approach.

The need for placement of at least three chimneys in the involved renal arteries and superior mesenteric artery to create a sufficient new sealing zone as in this case is associated with a remarkable risk of persistent gutters and technical difficulties, such as bilateral approach of the upper extremity. Two sheaths should be advanced from the left and one from the right side.[7] This condition can also provoke a cerebrovascular event. It is the outcome of the use of chimneys for treatment of aneurysms with endograft that remains unknown.[8,9]

Consequently, we decided to treat the patient by surgical means. A midline incision and lateral bowel displacement allowed rapid recognition and exposure of the aorta. After clamping of the aorta proximal and distal

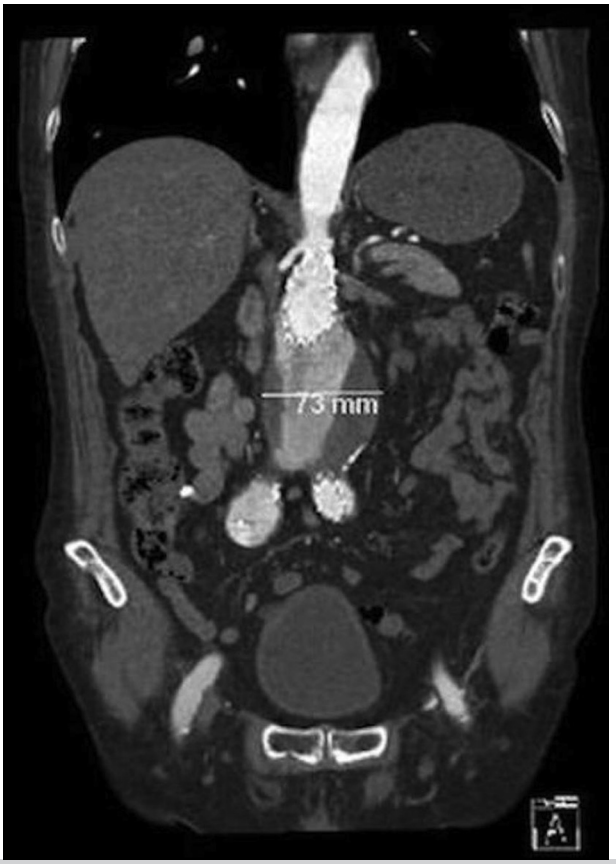

Fig 3. Computed tomography angiography revealed a 7.3-mm diameter of the aneurysm sac (coronal view).

to the iliac arteries, the stent graft was explanted in toto and aortic reconstruction was performed using a polyester graft.

The Achilles heel of EVAR remains the degeneration of the landing zones in patients with CTD causing type I endoleaks and necessitating reinterventions or open surgical conversion on the basis of the European Society for Vascular Surgery guidelines.[3] Waterman et al[10] reported 44% primary treatment failure after thoracic endovascular aortic repair or EVAR procedures in patients with Marfan syndrome, necessitating open conversion. The mortality rate for these patients after primary treatment with endovascular means was 43%.

Open repair appears to be a more efficient and suitable approach for this particular group of patients. Available articles related to CTDs are limited to the thoracic or thoracoabdominal aorta, showing encouraging results.[11] Experts, to succeed with a better approach in EDS patients' aorta and to decrease the risk of undesirable events (tears, rupture), proposed some specific techniques. These techniques include the use of intraluminal balloon catheters or external tourniquets to obtain circulatory control. In addition, soft peripheral arterial rather than heavy aortic clamps should be used to occlude the aorta. Conventional arterial clamps, even padded, atraumatic ones, tear when applied using the normal

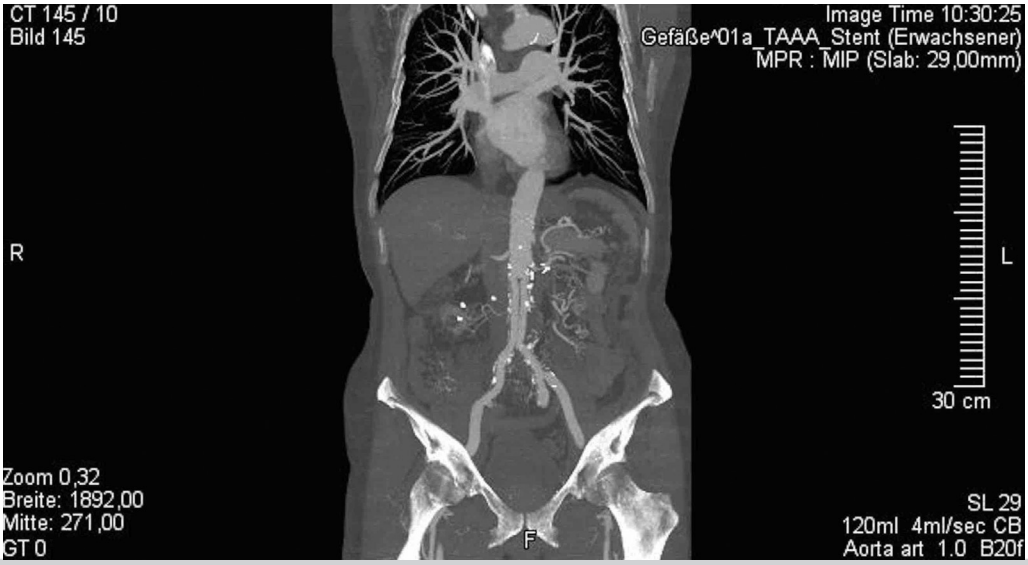

Fig 4. Computed tomography angiography revealed patency of the graft, free of other aneurysms and complications.

amount of force. Moreover, sutures may tear through the arteries, and this may be avoided by reinforcement with pledgeted Teflon and use of buttressed horizontal mattress sutures.[5]

Despite the challenging situation and the risk of life-threatening complications, promising outcomes, especially for patients with Marfan syndrome, have been published.[11] In expert hands, the technique seems feasible and durable with low reintervention and mortality rates.[11,12] Unfortunately, the literature remains devoid of EDS cases. Recently, a published study[11] did not clarify how many EDS patients died or suffered a complication during open repair. Based on antecedent studies,[13] the prognosis in type IV EDS patients may be poor, with an overall mortality rate of 63%; 19% die during the perioperative period.

In light of these recommendations and studies,[3,11,12] based on the continuing degenerative property of the disease, we attempted to give a more efficient and durable solution to this patient. Despite the paucity of patients with EDS and abdominal aortic aneurysms, this case reports the long-term follow-up of an EDS patient being treated by EVAR and highlights the limitation of the endovascular approach for this clinical entity.

REFERENCES

1. Brooke BS, Arnaoutakis G, McDonnell NB, Black JH 3rd. Contemporary management of vascular complications associated with Ehlers-Danlos syndrome. J Vasc Surg 2010;51:131-8; discussion: 138-9.
2. Eagleton MJ. Arterial complications of vascular Ehlers-Danlos syndrome. J Vasc Surg 2016;64:1869-80.
3. Riambau V, Bockler D, Brunkwall J, Cao P, Chiesa R, Coppi G, et al; ESVS Guidelines Committee. Editor's choice—management of descending thoracic aorta diseases: clinical practice guidelines of the European Society for Vascular Surgery (ESVS). Eur J Vasc Endovasc Surg 2017;53:4-52.
4. Svensson LG, Kouchoukos NT, Miller DC, Bavaria JE, Coselli JS, Curi MA, et al. Expert consensus document on the treatment of descending thoracic aortic disease using endovascular stent-grafts. Ann Thorac Surg 2008;85(Suppl):S1-41.
5. Bade MA, Queral LA, Mukherjee D, Kong LS. Endovascular abdominal aortic aneurysm repair in a patient with Ehlers-Danlos syndrome. J Vasc Surg 2007;46:360-2.
6. Pepin M, Schwarze U, Superti-Furga A, Byers PH. Clinical and genetic features of Ehlers-Danlos syndrome type IV, the vascular type. N Engl J Med 2000;342:673-80.
7. Donas KP, Criado FJ, Torsello G, Veith FJ, Minion DJ; PERICLES Registry Collaborators. Classification of chimney EVAR-related endoleaks: insights from the PERICLES Registry. J Endovasc Ther 2017;24:72-4.
8. Scali ST, Beck AW, Torsello G, Lachat M, Kubilis P, Veith FJ, et al. Identification of optimal device combinations for the chimney endovascular aneurysm repair technique within the PERICLES registry. J Vasc Surg 2018;68:24-35.
9. Donas KP, Torsello GB, Piccoli G, Pitoulias GA, Torsello GF, Bisdas T, et al. The PROTAGORAS study to evaluate the performance of the Endurant stent graft for patients with pararenal pathologic processes treated by the chimney/snorkel endovascular technique. J Vasc Surg 2016;63:1-7.
10. Waterman AL, Feezor RJ, Lee WA, Hess PJ, Beaver TM, Martin TD, et al. Endovascular treatment of acute and chronic aortic pathology in patients with Marfan syndrome. J Vasc Surg 2012;55:1234-40; discussion: 1240-1.
11. Keschenau PR, Kotelis D, Bisschop J, Barbati ME, Grommes J, Mees B, et al. Editor's choice—open thoracic and thoracoabdominal aortic repair in patients with connective tissue disease. Eur J Vasc Endovasc Surg 2017;54:588-96.
12. Coselli JS, Green SY, Price MD, Hash JA, Ouyang Y, Volguina IV, et al. Results of open surgical repair in patients with Marfan syndrome and distal aortic dissection. Ann Thorac Surg 2016;101:2193-201.
13. Karkos CD, Prasad V, Mukhopadhyay U, Thomson GJ, Hearn AR. Rupture of the abdominal aorta in patients with Ehlers-Danlos syndrome. Ann Vasc Surg 2000;14:274-7.

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