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Case report

Survival after spontaneous aortic rupture in a patient with Ehlers–Danlos syndrome

Camille Dambrin[a,*], Bertrand Marcheix[a], Tudor Bîrsan[b], Marie Bernadette Delisle[c]

[a]*Department of Cardiovascular Surgery (Pr Cerene), Rangueil Hospital, 1 Av Jean Poulhès, 31403 Toulouse Cedex 4, France*
[b]*Department of Surgery, Vienna General Hospital, Vienna, Austria*
[c]*Department of Pathology, Rangueil Hospital, Toulouse, France*

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Abstract

Ehlers–Danlos syndrome (EDS) is a rare inherited disorder of connective tissue characterized by hyperextensible skin, hypermobile joints, and abnormalities of the cardiovascular system. Most patients are unaware of their disease until a catastrophic event such as arterial rupture or bowel perforation occurs. Aortic disruption accounts for many of the deaths in EDS type IV cases and only two cases of survival after spontaneous aortic rupture have previously been reported. We report on a third case of a survivor of spontaneous abdominal aortic rupture in EDS type IV.

Keywords: Ehlers–Danlos; Aorta; Dissection; Vascular graft

1. Report

A previously healthy 25-year-old man presented in our department with spontaneous hematoma of the right thigh. The patient was hemodynamically stable. Vascular ultrasound showed a dissection of the right iliac artery with extension to the femoral artery. Because the proximal extension of the dissection could not be accurately seen on ultrasound, a computed tomography of the abdomen and pelvis was performed. This showed a dissection of the terminal abdominal aorta (Fig. 1a) extending to the right common iliac artery (Fig. 1b). We decided to treat surgically. In the meanwhile, the patient had also developed a collapse (BP: 70/30 mmHg, HR: 130/min) and was presenting a slightly distended and painful abdomen.

Upon performing a midline incision, an important retroperitoneal hematoma was found. The hematoma originated from a large tear in the infrarenal aorta. Bleeding was initially controlled by clamping the infrarenal aorta. The vascular tissue appeared to be extremely fragile and an additional tear in the aorta was made by the clamp. Therefore, the aorta was clamped by using small bulldog clamps. The right common iliac artery was dissected up to the right common femoral artery. A tear in the right common femoral artery close to its bifurcation was found, which was probably responsible for the spontaneous hematoma of the leg. Bypass grafting was performed using a 14 mm bifurcation graft (Dialine®II, BARD, France) which was sutured end to end to the aorta. The right limb of the graft was anastomosed end to end with the right common femoral artery. The left limb of the bifurcation graft was anastomosed end to end with the left common iliac artery. All anastomoses were performed by using 5/0 monofilament sutures (Prolene® ETHICON, France) placed in a horizontal interrupted manner and reinforced with felt pledgets. All sutures were further sealed with biological glue (Bioglue® Cryolife International, USA). Finally, the longitudinal injury of the aorta made by the clamp was repaired using 6/0 Prolene mattress sutures. The patient lost a total of 12,000 mL of blood and received 15 units of packed red blood cells, 10 platelet units, and 6 units of fresh frozen plasma. He was extubated 5 h after surgery and remained stable. He had an uneventful postoperative course and was discharged after 8 days. Two years later, our patient is doing well, and has had no problems since being in the hospital.

2. Discussion

In 1987, Cikritt reported 12 aortic perforations in EDS type IV patients [1]. The authors commented that all the ruptured aortic hemorrhages have been fatal. Few elective aortic reconstructions in case of EDS type IV have been reported [2–4]. Levine reported the case of a 14-year-old boy with EDS type IV who had aortic trauma. They managed to treat conservatively and the young patient survived [5].

* Corresponding author. Tel.: +33 5 61 32 26 52; fax: +33 5 61 32 23 15.
E-mail address: cdambrin@hotmail.com (C. Dambrin).

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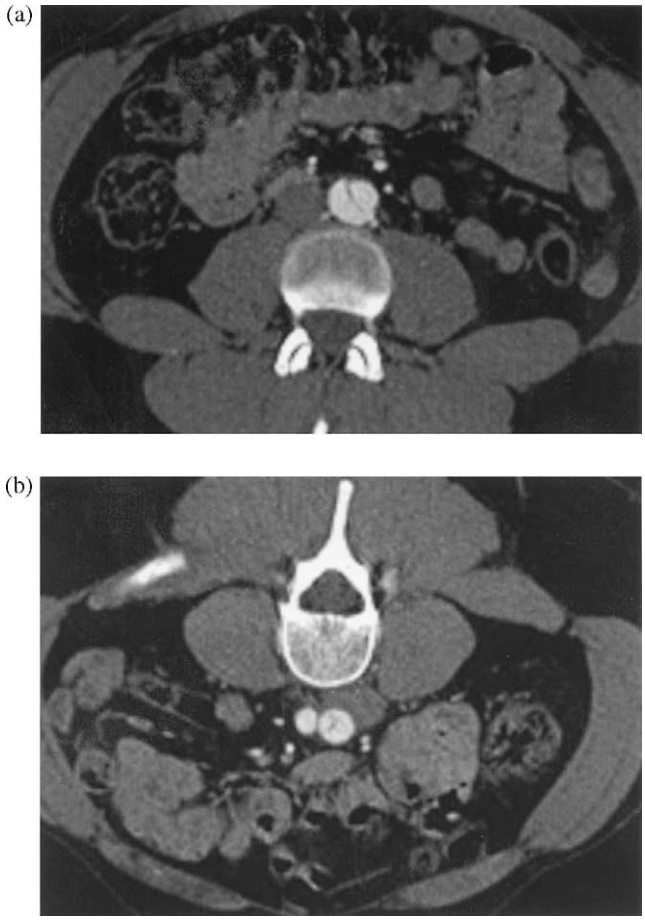

Fig. 1. (a) Computed tomography of the abdomen showing a dissection of the terminal abdominal aorta. (b) Computed tomography of the pelvis showing an extension of the aortic dissection to the right common iliac artery.

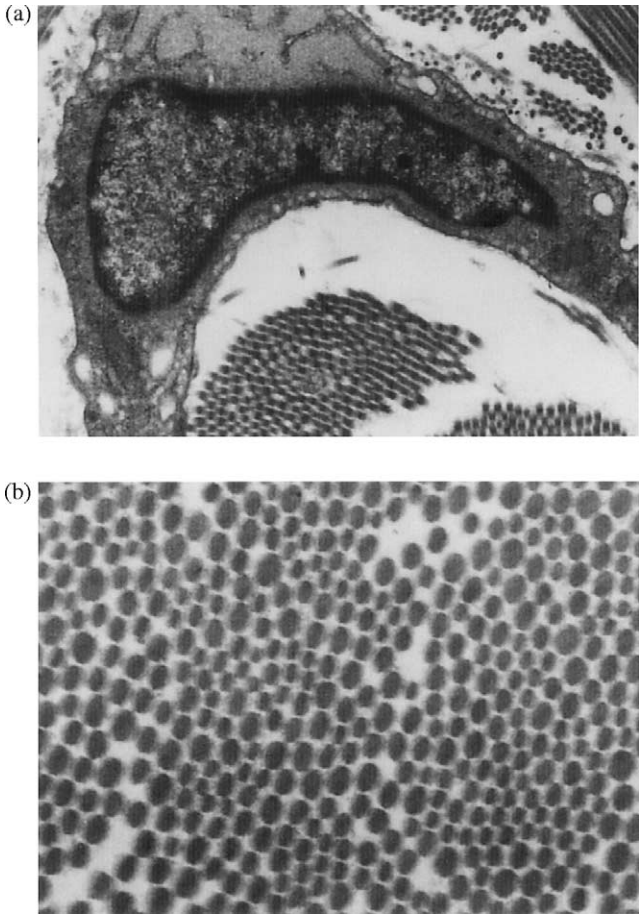

Fig. 2. (a) Electron microscopy demonstrating dilatation of granulous endoplasmic reticulum in dermal fibroblasts, a finding which is typical FOR EDS type IV. (b) Electron microscopy appearance of collagen bundles showing an irregularity of the contours and an abnormal range in the diameters of the round collagen fibers.

Only two survivors of spontaneous aortic rupture in EDS type IV patients have previously been reported. Gertsh reported the successful resection of a ruptured abdominal aortic aneurysm in a 52-year-old man with EDS type IV [6]. Serry reported a case of a 16-year-old boy who presented a spontaneous rupture of the abdominal aorta and was treated with an aortic bifurcation graft and survived [7].

Because experience with the operative technique in these patients is limited, the technical details presented may be of interest. The aortic clamps proved to be extremely dangerous instruments. Applied with normal force, they provided initial bleeding control but then caused a tear in the aortic wall and led to further bleeding. Vessel loops and internal balloon control have been advised by others. In this case, we have used small bulldog clamps for the abdominal aorta and vessel loops for the branches of the femoral artery. Aortic replacement has been achieved by the use of fine monofilament sutures placed in a horizontal interrupted manner and reinforced with synthetic pledgets. This seems to be the preferred technique for vascular repairs in true EDS type IV patients [5]. Any shear forces were avoided while performing the sutures. Local compression and the use of biological glue helped to achieve hemostasis.

Our patient had no family history suggestive for EDS. He had been previously healthy and had no obvious phenotypic features such as translucent skin or hypermobility of joints. However, his young age as well as the clinical presentation were peculiar and should have let us suspect a connective tissue disorder. Postoperatively, careful examination showed a scar on the knee of which the aspect was typical of EDS.

The diagnosis of EDS type IV was confirmed post-operatively in a skin biopsy specimen which revealed a lack of type III collagen. The electronic microscopy examination revealed a typical aspect of EDS type IV (Fig. 2a,b). To our knowledge, this report is the third documented survivor of a spontaneous aortic hemorrhage in EDS type IV.

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