

Successful Coil Embolization for Spontaneous Arterial Rupture in Association with Ehlers-Danlos Syndrome Type IV: Report of a Case

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Abstract

When a patient with Ehlers-Danlos syndrome (EDS) presents with a vascular emergency, performing life-saving surgery can be difficult because of the profound fragility of the arterial tissue. We report the case of a 27-year-old woman with EDS in whom a spontaneous arterial rupture was successfully treated with transcatheter embolization. The patient was brought to our hospital in shock, with left lower abdominal pain. She had been diagnosed with EDS type IV following a colonic rupture 8 years earlier. An emergency angiogram revealed rupture of the left external iliac artery. The active bleeding was managed by transarterial embolization of the ruptured artery using stainless steel coils, which took 30 min to achieve. The patient has not suffered any further vascular complications during the year since this procedure. Transcatheter coil embolization may be a reliable option for treating sudden arterial rupture in patients with this syndrome.

Key words Ehlers-Danlos syndrome · Spontaneous arterial rupture · Coil embolization

Introduction

Ehlers-Danlos syndrome (EDS) is an uncommon inherited disease of connective tissue.^{1,2} It has been classified into ten types according to its clinical manifestations. Type IV EDS, the vascular type, is of particular concern to surgeons because it is frequently associated with catastrophic complications such as spontaneous arterial or colonic rupture. In fact, 44% of type IV EDS patients with arterial rupture die before any surgical intervention can be instituted and 19% die during or immedi-

ately after the operation, resulting in an overall mortality of 63%.³ Although potent strategies are needed for the emergency management of this vascular complication, performing vascular surgery on patients with this syndrome is difficult because the arterial tissue is so fragile.³⁻⁵

We successfully treated a spontaneous arterial rupture in a patient with EDS by performing transcatheter embolization. The usefulness of this modality in the management of EDS-related vascular emergencies is discussed.

Case Report

A 27-year-old woman presented to our hospital with left lower abdominal pain and signs of shock. On admission, her blood pressure was 80/40 mmHg, heart rate 120 bpm, and hemoglobin 9.0 g/dl. Her skin was thin with visible veins and swelling was seen in her left lower abdomen. She had a colostomy stoma after undergoing a sigmoid colostomy at another hospital for sigmoid colonic perforation 8 years earlier, at which time a clinical diagnosis of EDS type IV had been made. There was no known family history of EDS.

Spontaneous arterial rupture of the left iliac artery was suspected based on computed tomographic findings, which revealed a large hematoma in the retroperitoneal space, in the left lower abdomen (Fig. 1). An emergency angiogram showed that the aorta and the iliac artery were small in diameter and that contrast media appeared to be leaking from the left external iliac artery, indicating an arterial rupture (Fig. 2). It was necessary to transport the patient to perform surgery. Exposure of the iliac artery was considered to be relatively difficult because of her history of abdominal surgery. Therefore, we decided to control the hemorrhage by performing transcatheter coil embolization of the ruptured artery.

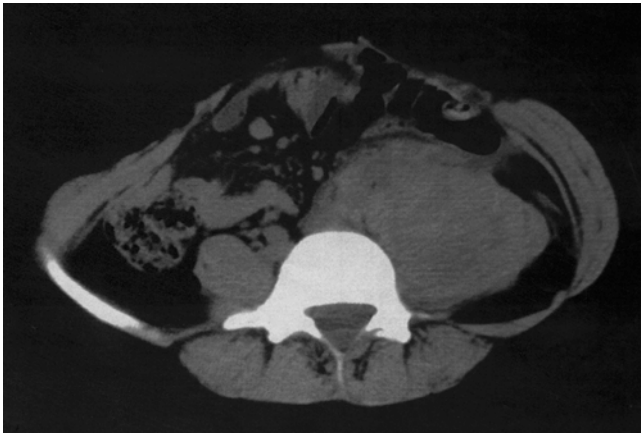


Fig. 1. Computed tomographic scan revealed a large hematoma in the retroperitoneal space of the left lower abdomen

The aorta was occluded just proximal to the bifurcation to enable precise positioning of the coils. An occlusion balloon catheter was inserted through the right femoral artery, and Gianturco stainless steel coils (Cook, Bloomington, IN, USA) were introduced into the left external iliac artery via the left femoral artery. A total of eight coils, six of which were 8 mm in diameter and 5 cm in length, and two, 6 mm in diameter and 5 cm in length, were needed to complete the embolization. Following the procedure, the left femoral artery was found to be well perfused via the collateral arteries arising from the left internal iliac artery (Fig. 3). The patient had no symptoms of lower limb ischemia, and surgical revascularization was not required.

The patient had an uneventful clinical course. No problems arose at the entry site of the artery. She was discharged 10 days after treatment and has not experienced any further vascular complications in 1 year of follow-up.

Discussion

The etiology of EDS type IV involves a mutation in the gene for type III procollagen.² This defect is diagnosed on the basis of four clinical criteria; namely, easy bruising, thin skin with visible veins, characteristic facial features, and rupture of the arteries, uterus, or intestine. It was reported that 25% of patients in this type of EDS have an initial event necessitating surgical intervention before the age of 20 years, and more than 80% suffer at least one complication by the age of 40.² Our patient had undergone the surgery for the first time following colonic rupture at 19 years of age. Although the diagnosis was not confirmed by laboratory findings of genetic and dermal fibroblast studies, the manifestation of

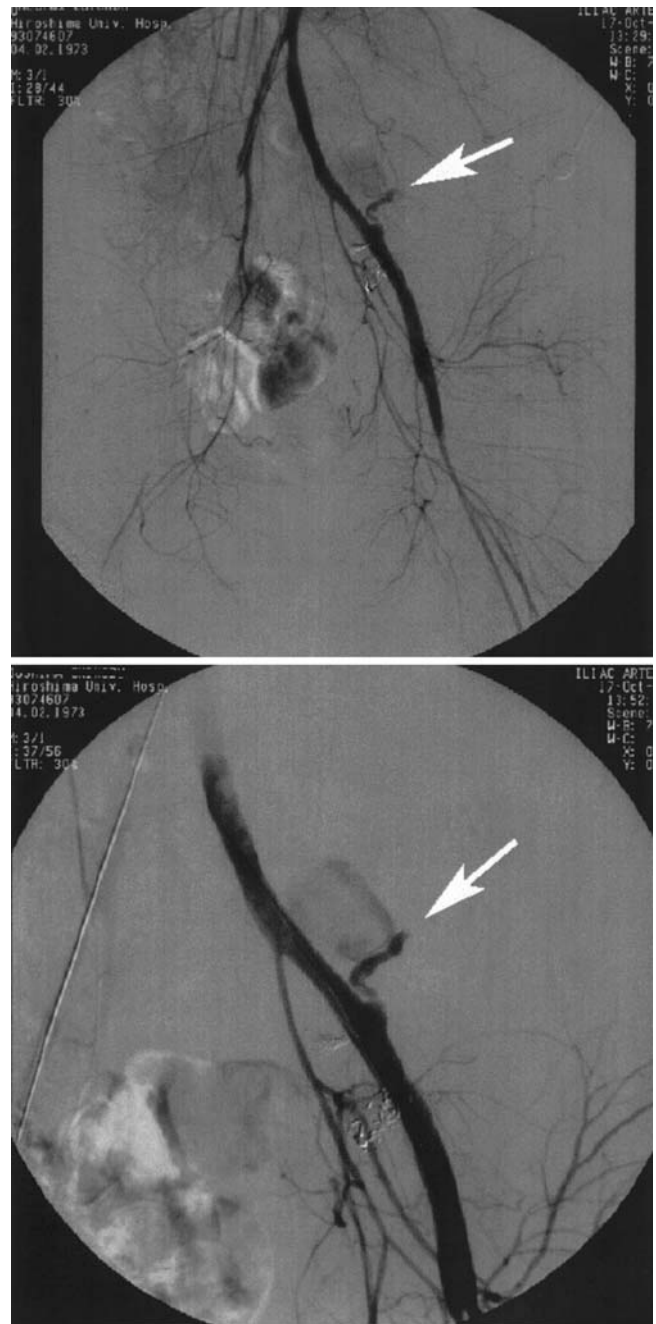


Fig. 2. Angiogram before embolization. Contrast medium was seen leaking from the left external iliac artery (arrows)

two uncommon complications justified the clinical diagnosis.

Most patients with this type of EDS die of a spontaneous arterial rupture, the median survival being 48 years.² Moreover, the treatment of this complication must be conservative because cross-clamping can easily cause an arterial tear, resulting in uncontrollable bleeding.³⁻⁵ Thus, endovascular methods may be advantageous for the management of this catastrophe, because



Fig. 3. Angiogram after embolization. The *arrow* indicates the coils used for embolization

minimal manipulation of the fragile arteries is required and they can be carried out immediately after angiographic determination of the rupture site. Currently, stent grafting and transcatheter embolization are the most feasible treatments of arterial rupture. However, the application of a stent graft in patients with this vasculopathy is associated with two potential problems. First, the stent graft can injure the arterial wall in the area where it has been placed. Hovsepian et al.⁶ described a pseudoaneurysm at the proximal edge of a stent graft that had been deployed to cover the mouth of an axillary pseudoaneurysm in a patient with EDS. Second, it is difficult to procure a stent graft suitable for vascular application in an emergency situation because they are not commercially available in Japan.

On the other hand, coil embolization is generally used to control active hemorrhage, such as that caused by arterial trauma secondary to pelvic fractures.⁷ This method has been proven to effectively embolize arteries as large as the iliac vessels.⁸ Needless to say, the transcatheter embolization of arteries perfusing an essential organ is controversial. However, when this technique is applied to limb arteries, consequent revascularization must be considered for limb rescue. If our patient had suffered lower limb ischemia after intervention, a femoral crossover bypass would have been the optimal choice.

In conclusion, this minimally invasive modality may be a therapeutic option for vascular emergencies in patients with EDS type IV. However, intensive follow-up of treated arteries is mandatory because coil placement potentiates injury to the walls of EDS-affected vessels.

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