

# Endovascular Treatment for a Spontaneous Rupture of the Posterior Tibial Artery in a Patient with Ehlers–Danlos Syndrome Type IV: Report of a Case

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## Abstract

This report presents the case of a 27-year-old woman with a history of Ehlers–Danlos syndrome Type IV (EDS-IV) who suffered from acute compartment syndrome caused by the spontaneous rupture of the posterior tibial artery. Bleeding control was achieved by endovascular embolization. A fasciotomy was successfully performed without any trouble with hemorrhaging. Endovascular treatment could therefore represent the ideal option for treating infrapopliteal lesions, particularly for EDS-IV patients.

**Key words** Ehlers–Danlos syndrome · Compartment syndrome · Spontaneous rupture

## Introduction

Vascular complications of Ehlers–Danlos syndrome (EDS) type IV (EDS-IV) have been reported to be a fatal condition in which vascular surgeons need to consider more sophisticated approaches. In one case series, more than 70% of EDS-IV patients developed vascular complications and cause of death was presumed to be associated with vascular complications in almost all cases.<sup>1</sup> In bleeding patients, either an emergency surgical repair or an endovascular approach must be selected depending on various factors.<sup>2</sup> Comorbid surgical situations such as acute compartment syndrome of the limb complicate the treatment strategies. Acute compartment syndrome of the leg caused by a spontaneous arterial rupture in an EDS-IV patient has not been previously reported.

## Case Report

A 27-year-old woman with a history of EDS-IV was brought to the emergency room complaining of a swollen and painful right leg. She had a history of spontaneous pneumothorax at 21 years of age and a diagnosis of EDS-IV was made at that time. One week before admission, she felt pain in the right leg when playing badminton. She also noticed that the right leg was swelling, but those symptoms later improved spontaneously. On the morning of her admission, the leg pain suddenly became exacerbated. The pain had been worsening and she was unable to walk unaided. Her initial blood pressure was 120/80 mmHg, heart rate 90 beats/min, and respiration rate 20 breaths/min. The patient appeared quite distressed. Prominent swelling was apparent in the right leg and subcutaneous hemorrhage was also evident (Fig. 1). Her distal pulse was palpable but passive dorsal movement of the right ankle caused pain in the posterior aspect of the leg. Her initial leukocyte count was 11 200/ $\mu$ l and hematocrit was 35.6%. Computed tomography (CT) of the lower extremity showed the formation of a posterior tibial arterial aneurysm and extravasation of contrast material, thus leading to a diagnosis of compartment syndrome caused by spontaneous rupture of a posterior tibial artery (Fig. 2). Angiography revealed a posterior tibial artery aneurysm about 2 cm in diameter with leakage of intravenous contrast (Fig. 3A). A microcatheter was inserted through the contralateral femoral artery. Tornado embolization microcoils (Cook, Bloomington, IN, USA) and VorTX-18 Diamond shape microcoils (Boston Scientific, Natick, MA, USA) were used to complete the embolization of the proximal and distal sides of the aneurysm. Postembolization angiography did not show any flow into the aneurysm (Fig. 3B). No ischemic signs were seen in the right lower extremity after embolization. The patient was then brought to the operating room and underwent a fasciotomy of the right leg. Preoperative measure-

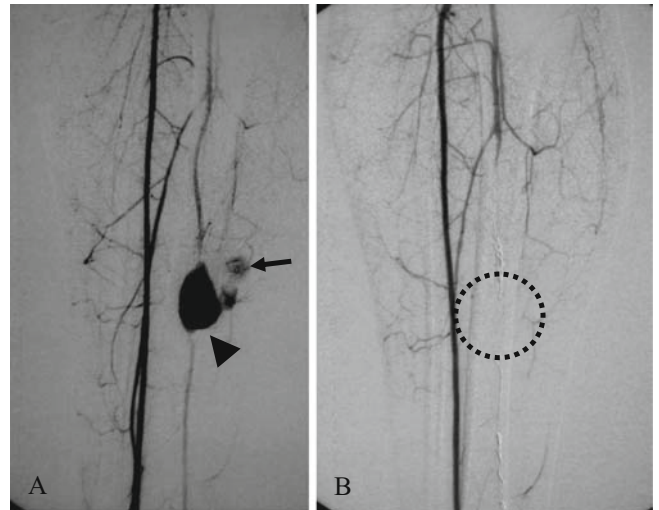


**Fig. 1.** Initial findings in the right leg. Swelling and purpura were grossly abundant in the right leg

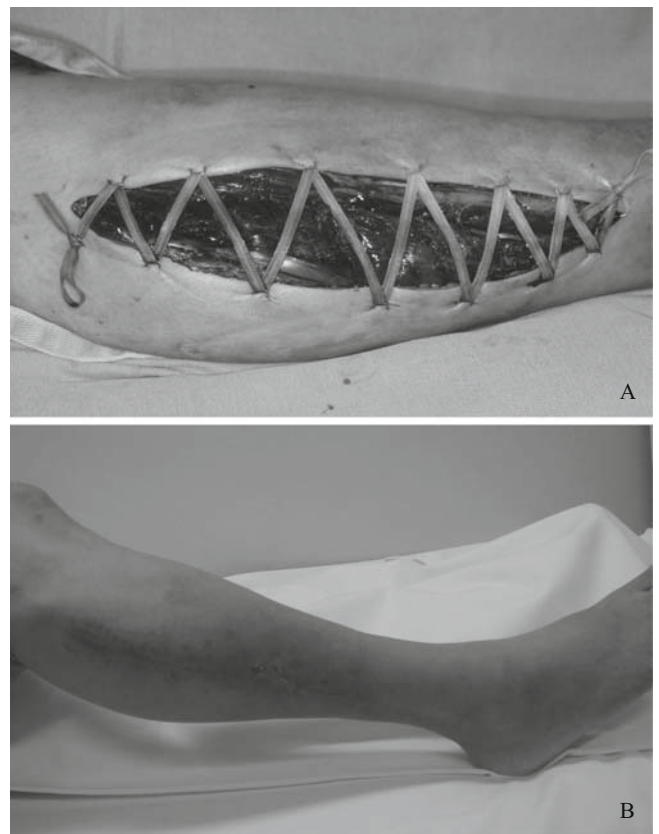


**Fig. 2.** Coronal (A) and sagittal (B) reconstructions of contrast-enhanced computed tomography showed a posterior tibial arterial aneurysm (arrows) with extravasation of the contrast medium (arrowhead)

ments of the intracompartmental pressure were 31 mmHg, 19 mmHg, 38 mmHg, and 45 mmHg in the anterior, lateral, superficial posterior, and deep posterior compartments, respectively. All four compartments were decompressed by a longitudinal incision along the lateral leg. No difficulties were encountered in controlling hemorrhaging during the surgery. The wound was kept open using a shoelace technique, in which a vessel tapes were crossed repeatedly from one skin edge to the other edge under skin staples, enabling approximation of skin edges day by day (Fig. 4A). Finally, it was possible to close the wound without a skin graft and the



**Fig. 3.** A Emergency angiography of the right leg identified a posterior tibial artery aneurysm (arrowhead) and extravasation (arrow). B Postembolization angiography confirms the excluded posterior tibial artery (dotted circle)



**Fig. 4.** A Postoperative wound of the right leg. Sterile bands are stapled to the skin edges over the fasciotomy site. B After the skin edges were approximated, the wound was closed successfully without a skin graft

patient recovered successfully without any functional problems (Fig. 4B).

## Discussion

Ehlers–Danlos syndrome is a rare inherited disorder that affects the connective tissue. In 1997, the classification of EDS was revised to six types, including classical, hypermobility, vascular, kyphoscoliosis, arthrochalasis, and dermatosparaxis type.<sup>3</sup> Ehlers–Danlos syndrome type IV, vascular type, is an autosomal dominant disorder resulting from mutations in the gene for type III procollagen (COL3A1).<sup>4</sup> The clinical diagnosis of EDS-IV is made on the basis of four clinical criteria: easy bruising; thin skin with visible veins; characteristic facies; and rupture of arteries, uterus, or intestines.<sup>3</sup> This patient had a family history of EDS-IV and satisfied all the criteria of diagnosis. Due to the fragility of the vessels, a well-accepted strategy for dealing with vascular complications of EDS-IV is a conservative approach, with surgical treatment reserved for patients presenting with imminent or frank life-threatening bleeding.<sup>1</sup>

Compartment syndrome is a condition characterized by an increased pressure within a closed space, with the potential to cause irreversible damage to the compartment contents. This syndrome is commonly described in the leg or forearm, but can affect the thigh, hand, or other areas of the extremities.<sup>5,6</sup> The etiology of compartment syndrome can be categorized as: (1) increased compartment contents; (2) decreased compartment volume; and (3) external pressure. Although trauma patients suffering from a fracture or deep burns are frequent victims of compartment syndrome of the leg, the formation or rupture of aneurysms has rarely been noted as a cause, especially in the infrapopliteal arteries.<sup>7</sup> Patients with compartment syndrome usually complain of severe pain in the extremity. A physical examination reveals that the patients may have tension and tenderness of the affected compartment, passive motion pain of muscle in the compartment, sensory disturbance, or weakness of the extremity.<sup>8</sup> The diagnosis of compartment syndrome is confirmed by the measurement of tissue pressure. A decompressive fasciotomy is the primary treatment to normalize elevated compartment pressure. Although controversy remains regarding which compartment pressure should be an indication of a fasciotomy, a compartment pressure in the range of 30–35 mmHg is often considered to be the threshold. A delay in performing surgery can lead to irreversible muscle and nerve damage. Once compartment syndrome is diagnosed, surgery must be performed as quickly as possible.

An aneurysm of the infrapopliteal artery is a rare vascular lesion. The common etiologies of a pseudoan-

eurysm include blunt or penetrating trauma and transluminal angioplasty, leading to a full-thickness tear or lacerations of the vessel wall.<sup>9–12</sup> On the other hand, true aneurysms of the infrapopliteal artery are usually diagnosed as mycotic.<sup>13</sup> The pathophysiology in the current case may have involved an initial spontaneous rupture of the posterior tibial artery that occurred when the patient played badminton, which thereafter progressed to a pseudoaneurysm. On the day of admission, the rupture of the pseudoaneurysm caused the compartment syndrome of the leg. Generally, the treatment options for an aneurysm in this area are: (1) excision of aneurysm followed by reconstruction; (2) endovascular embolization; (3) ligation of the proximal and distal arteries; and (4) percutaneous occlusion by means of fibrin adhesive injection.<sup>9</sup> Recently, there have been an increasing number of reports of successfully treated crural artery aneurysm.<sup>12,14,15</sup> The advantages of the endovascular approach are the protection of the surrounding nerves and adjacent structures during repair and the minimal blood loss. Conversely, the disadvantages include the risk of thrombosing the narrow supplying artery either by stent-graft implantation or by transcatheter introduction of coils and a higher rate of failure.<sup>11</sup> At present, there is no clear consensus on the optimal strategy for infrapopliteal aneurysm. In cases of a ruptured aneurysm complicated by acute compartment syndrome, an emergency fasciotomy and subsequent surgical treatment for the aneurysm is often selected, but the story is completely different in cases of EDS. The recommended strategy in dealing with vascular complications of EDS-IV is a more conservative approach because of the excessive tissue fragility, which often causes catastrophic complications.<sup>1,6,16</sup> Furthermore, since conventional arteriography has been associated with a 67% complication rate and a 17% mortality rate, meticulous endovascular techniques are required for handling the catheter.<sup>6</sup> In the present case, endovascular embolization was performed first. The risk of uncontrollable bleeding associated with a fasciotomy took precedence over the hemostatic procedures. As a result, a decompressive fasciotomy was successfully performed in a dry operative field after embolization. In addition, surgeons need to take the diagnosis of EDS into account before surgical intervention. Sophisticated approaches including endovascular techniques should therefore be used in cases of concurrent mandatory surgery in patients with EDS.

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